

# ***ALS From Both Sides***

Care of an ALS Patient

By Diane Huberty, Retired RN, Certified Neuro Nurse...and ALS Patient

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As someone amazed to find herself in her 30th year of ALS and still busily, happily, and contentedly engaged in living, I have reached the point where I find myself saying "If I'd known I would live this long, I would have taken better care of myself!" As a nurse specialist in Neuroscience, I knew the long term problems of immobility and patient care issues. But with a diagnosis of ALS, I wasn't thinking long term. Well, here I am, big time long-term! I find myself looking at ALS from both sides, as a nurse, and as an ALS patient. For that reason, one focus of this site is dealing with some of the medical complications we face as a result of ALS. All too often these things are inadequately addressed because the expectation is we will not live long enough to worry about "long term" problems. These are problems that ALS patients and caregivers should anticipate, recognize, and minimize in order to assure a good quality of life even if that life is short. This is increasingly important because several factors are likely to extend the life of ALS patients in years to come:

- The development of medications to slow progression.
- Continued improvements in supportive care (nutrition, respiratory support etc.)
- Computer aided communication and environmental control equipment to greatly improve the quality of life for ALS patients who opt for ventilation. As a result more of us will take that option and live for many more years.

In addition, as someone with ALS, I have found that medical problems are only half the battle. Dealing with the practical issues of day by day ALS patient care and adaptation to increasing disability is as much a part of the quality of life as medical problems. In my years of slow progression, I have gone through (and continue to go through) the many stages of ALS. Slow progression has meant that I have spent years instead of months in each stage and have had time to problem solve the hassles of daily life. I hope that some of these ideas will turn what was a lot of trial and error for me into a fix that even those with rapid progression can use.

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***Diane Huberty, January 31, 2018  
dianehuberty@frontier.com***

## ***Muscle Cramping and Spasticity***

There are four main "muscle things" that occur with ALS: Spasticity, cramping, fasciculations, and fibrillations.

### *Muscle Cramps*

Muscle cramps are very common in ALS, probably due to muscle fatigue or irritability from screwed up nerve impulses. They can be terribly painful and can occur in any muscle; feet, legs, arms, abdomen, chest, back, and (my all time favorite for sheer pain!) the neck and jaw. They can be in small muscles or affect big ones -- good old-fashioned charlie horses. As the muscles are affected by ALS, cramping is noted in that area. The cramping becomes less severe with time because the weakening muscles simply can't work up a good cramp anymore. Cramps tend to be repetitive. Once a muscle starts cramping up, it does so over and over for a miserable hour or so. They tend to occur more if you have overdone exercise, if the muscle is cold, or if circulation is decreased. Holding a book up to read in bed on a cool night will just about guarantee some whopping good hand cramps and result in interesting finger gestures!

### *Spasticity*

Spasticity is an upper motor neuron problem and is present to some degree in ALS. For some patients, it is minimal, for others extreme. Spasticity can actually be helpful in maintaining function as the rigidity helps replace normal muscle strength, but it causes jerky, hard to control movements. Spasticity causes a tightening of muscles that results in a stiffening of that part of the body in an exaggerated reflex. It is actually triggering both the muscles to flex and the muscles to extend that part of the body at the same time. It can occur in any muscles -- the arms, legs, back, abdomen, neck, or entire body at once. A simple touch can trigger it and it may last only a moment or persist indefinitely. Spasticity isn't always painful but it can be, especially if it triggers muscle cramps that add to the pain.

### *Fasciculations*

Fasciculations (muscle twitches) are probably due to nerve irritability. They occur in smaller muscle bundles inside large muscle bundles and can be observed as well as felt. Fasciculations are not so much painful as irritating. It can feel as if someone is popping corn under your skin! They can be incredibly persistent and keep you from sleeping.

### *Fibrillations*

Fibrillations occur in single muscle fibers within a muscle. They cannot be felt but can be seen on EMG (Electromyogram). Fibrillations occur in conditions besides ALS.

### *Is it Spasticity or Cramping?*

The first step in relieving these recurring pains is determining whether they are due to spasticity or cramping. Medications that work for spasticity don't necessarily work for muscle cramps and vice-versa.

Spasticity affects larger areas of the body -- arm, leg, trunk, neck. All the muscles in the area tighten up and the entire area may be so tight it hurts. Muscle cramps are generally limited to a single muscle and the pain, although intense even in small muscles, is limited in area.

Spasticity often begins with an odd sensation, sometimes described as a quiver, rushing and spreading through the area and becoming painful as they tighten the muscles and more painful as the muscles tire. Muscle cramps start out painful and just get worse as they tighten up.

Spasticity in the legs generally causes the legs to stiffen out. In the trunk, back or neck it causes your body to arch a bit. In the arms, it may flex or extend the arm. Spasticity is often most striking when you first wake up or start to move after sitting still for a while and can often be connected to a sensory trigger. That trigger can be as mild as a gentle touch so the connection may not be made.

Muscle cramps often affect the hands and feet making fingers and toes curl incredibly tightly. Spasticity is more widespread, less likely to affect just those muscles, and more likely to stiffen them out than to curl them.

### *Medications*

Spasticity can usually be helped by medication but can be a very stubborn problem. One consideration in treating spasticity is to find a balance between relieving excessive and painful spasticity and maintaining a certain level of spasticity which can be helpful by replacing muscle strength.

The meds for spasticity are primarily Baclofen, Dantrium and Zanaflex, tizanadine HCL.

Although Baclofen is often listed under meds for cramping and prescribed for it, the indications say "Clinically, Baclofen is used to treat spasticity." In my experience, Baclofen did not reduce cramping at all. However, if spasticity is triggering the cramping, Baclofen may reduce cramping by reducing the spasticity. Some patients experience weakness or tiredness while taking it but these problems can often be reduced if the dosage is decreased. In 1996 the FDA approved the use of Baclofen, delivered by an implanted pump, for the treatment of spasticity due to spinal cord injury and this is now being used for ALS and PLS patients.

Zanaflex is less likely to cause weakness but may cause sleepiness. This can often be minimized by starting with a low dose and gradually increasing it until spasticity is relieved.

Dantrium has been used for spasticity, but because of its potential for causing liver problems, is less frequently used since other meds are now available. It does seem to still be used for bladder control problems caused by spasticity, however.

The meds for cramping include:

Valium and other muscle relaxants may be used for muscle cramps. However, their side effects and cost make them a second choice to quinine in my opinion.

\*\*\*Update. Mexilitine is gaining in use, but like Quinine has possible effects on heart rhythm.

Quinine (Quaalun, Quinine Sulfate) is a drug long used for muscle cramping but in 1995 the FDA made it available by prescription only because of its very rare but potentially severe risks (heart arrhythmias). Even so, I would certainly recommend asking your doctor about trying quinine. Additionally, the FDA removed muscle cramping from its list of uses because leg cramps are not life threatening and therefore not worth the potential risk of quinine. Making matters worse for ALS patients who experience severe muscle cramps daily, the FDA limited quinine to a single formulation available only as Quaalun -- which costs around \$5 a tablet! All of this was done and continues in effect even though research shows quinine to be effective against cramping, and that

no other drug is significantly effective. #\*%&! So we are left to suffer or find a doctor willing to order Quaalun outside of the FDA listed use, and pray that insurance will cover it. If you find a doctor willing to write a prescription for generic quinine sulfate, save money by having the prescription filled by an Internet Canadian pharmacy. Or drink Tonic Water. The amount of quinine is probably too low to have any effect, however.

I do not recall what dose I took, but do know that too large a dose will cause weakness. I am told the smallest tablet made is 260mg. but I would recommend beginning with half a tablet and increasing to a whole one if it is ineffective. It is also available in 200 mg capsules. It is much less expensive than the other drugs prescribed, and, in my experience, the most effective.

Like any drug, quinine does have some potential side effects (and cannot be taken by pregnant women) but it does not cause physical and psychological dependence as Valium does in long term use. Aside from the weakness when I took the larger dose, I noticed no other immediate side effects. After more than 5 years of daily use, I did notice sweet foods started to lose any sweet taste, especially chocolate. Quinine is very bitter so I assumed it was the culprit since I wasn't on any other meds at the time. It was a minor problem and when I did stop taking quinine, I found the ability to fully enjoy an Oreo was gradually restored!

There are other medications and home remedies to try. Retigabine (Trobal) is mentioned as is a spoonful of French's mustard or pickle juice! Magnesium, potassium, calcium are just some things suggested but should be used as ordered by a doctor. Excesses of these can cause severe problems. More importantly, they are less likely to be effective to any definite degree unless your levels were low to begin with -- and that is not usual in ALS. In ALS muscle cramping is caused by nerve irritability and/or muscle strain as muscles weaken.

I have not heard of any consistent success in reducing the fasciculations (muscle twitching) with any medication.

## *Choking: Variations on a Theme*

It's not uncommon in ALS for something to unexpectedly and abruptly aggravate your airway and throw you into a choking fit. I know from personal experience that these choking, gagging, gasping spells are physically exhausting, frustrating and extremely unnerving for both the choker and the caregiver. The actions to take for choking depend on the type of choking spell you're having. For example:

### *The Heimlich Choke*

A large piece of food completely blocks the airway. You can't speak or cough. This is an emergency! It requires the Heimlich maneuver, either by a caregiver or self-administered, to force the blockage up and out. If not successful in two tries, call 911 and continue trying the Heimlich while waiting for help.

### *The Damn-Near Heimlich Choke*

A pill or bit of food sticks in your throat. After a moment you can breathe but with a scary "heeee" sound. Just as you start getting a little air in, you start coughing and/or gagging again. Over and over. You can feel the darn thing stuck in your throat teetering between sliding up and making you gag and sliding down and making you cough. This is scary, exhausting, but you can get a breath in between coughing spells. The Heimlich or assisted cough technique may help. Focus on getting the obstruction up and out, not down the throat, because of the danger that it will lodge more firmly or go into the lungs. Therefore, avoid back whacking!

### *The Foodless Sneaky Choke*

You're just sitting there minding your own business or even snoozing when suddenly you're coughing and choking. A simple swallow of saliva goes awry and sends your larynx (vocal cords) into a spasm that closes off your airway. This is called a laryngospasm and isn't uncommon in ALS. You fight to inhale but nothing happens. You can't take a breath, can't call for help, and panic crashes over you. After an eternity, you finally suck in a thin, wheezing, tiny "eeee" breath. Slowly the spasm eases.

The only thing I've found that helps me through these is the hard-learned lesson that they're intense but short-lived. You're not really choking, you can get some air, and the spasm will ease in just a moment or two. Caregivers can help by giving calm reassurance, gentle back rubbing (not whacking) and other support in riding out the spasm.

### *The Phlegm From Hell Choke*

You try to clear your throat. Hrrrmph. Hrrrrrrrmph. No good. Over and over you try to break up and cough out the thick clog in the back of your throat until you're worn out and gasping. My treatment is to take a dose of papaya juice or expectorant (guaifenesin, available over the counter) to thin the secretions. Then I lie down (BiPAP on) and have my caregiver do an assisted cough or use a cough assist machine. By lying down, I don't have to fight gravity to bring the mucus up. Staying well hydrated can help keep secretions thinner.

### *The Non-Choke Sticker*

A pill or bit of tortilla chip sticks on the way down and sits there feeling like a thorn in your throat. This little sucker can hurt for a day or more. After taking a small sip to make sure I can still swallow, I try taking the longest drink I can. Sometimes this repeated swallowing does the trick.

Advice to Caregivers: In any choking situation, fake being calm and quickly go through a list of possible actions. Ask, "Do you need a Heimlich? Assisted cough? A drink? Cough medicine? Suction?"

Besides the physical and emotional stress of a choking attack, the real danger of suffocation and the loss of calories due to fear of eating, choking can lead to aspiration, in which food or pills go into the lungs and cause life-threatening pneumonia.

Prevention is the key here. Go to *Swallowing*, Page 7.



## *Swallowing: A Risky Business*

Remember the old drinking toast "Over the lips and past the gums, look out stomach, here it comes!" A fun drinking salute but misguided. It isn't the stomach that needs to be on the lookout, it is the lungs that are at risk if anything goes wrong! Swallowing is a coordination of effort by a number of sensory and motor nerves in a series of reflexes, and a missed step has that food or drink headed for the lungs.

The swallowing process has three stages. The oral phase includes chewing the food using the tongue to move the food around and saliva moistening the food. Then the tongue lifts up in front and pushes the food to the back of the mouth. All this is voluntary movement and voluntary movement is what is affected by ALS. Tongue movement, chewing, and the ability to close the lips are all affected.

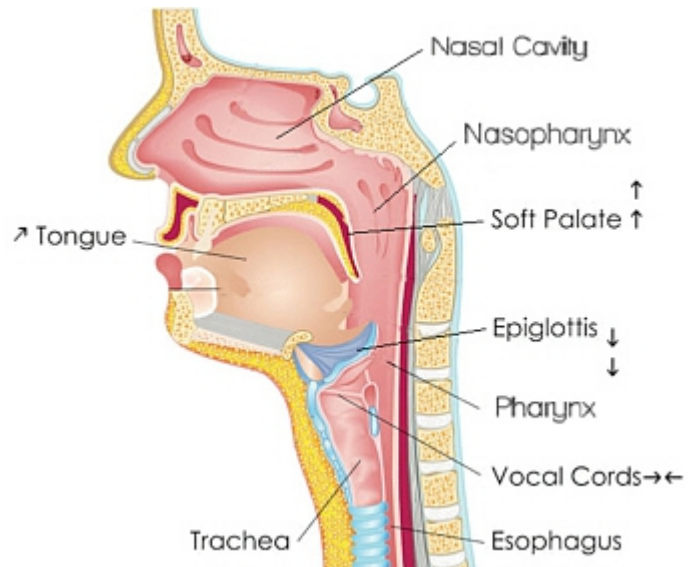
Usually however, the second phase of swallowing is affected first. The second phase of swallowing is a series of reflex movements we have no control over. Once the series is triggered by the food entering the top of the throat, the food moves down whether or not all the reflexes are in working order. The soft palate moves up to block food from going up into nasal passages.

(Everyone who has burst out laughing mid-swallow of a liquid knows that this reflex doesn't provide a strong seal!) The epiglottis begins the most important part of the swallow; keeping the food or drink from entering the trachea and lungs. The epiglottis is like a trap door that is reflexively triggered to drop down over the tracheal opening.

Just below it, the vocal cords squeeze together to form an even stronger barrier. With nowhere else to go, the food or drink is pushed down into the esophagus by the muscles of the pharynx, not pulled by gravity. The esophageal phase of swallowing moves the food or drink on down into the stomach.

All these reflexes require motor responses from cranial nerves, including V, VII, IX, X and XII. The cranial nerve centers are all in the brain stem, the connection of the brain to the spinal cord. Some anatomical wit thought the brain stem looked like the bulb of a plant, so we have the term "bulbar" to refer to anything going on in the brain stem, especially involving the cranial nerves. With ALS we may have bulbar onset. In addition to early signs of swallowing problems beginning with a simple cough when swallowing, a change in the tone and quality of the voice is often noted. Even without bulbar onset, ALS progresses to include bulbar problems. Choking on food or liquids begins intermittently and progresses to inevitable.

The immediately life threatening problem is choking badly enough to block the airway. A slower but inevitable and potentially fatal problem happens as the muscles needed to swallow safely weaken, and food and liquids are aspirated into the lungs causing pneumonia. When the swallowing muscles are no longer working it is possible to swallow without coughing because the



cough and gag reflexes are gone. This is called Silent Aspiration. The food or liquids go into the lungs with no cough to signal a problem.

As swallowing worsens, health and quality of life deteriorates. Dehydration causes low blood pressure, dry mouth, and adds to problems with lung congestion and constipation. Meals are long and drawn out from difficulty chewing, exhaustion, and frightening choking episodes. Both patient and caregiver dread meal times and fear that each choking episode may be the last. Food choices become limited, and eventually even blenderized food is dangerous. Weight loss is significant. If you persist in trying to get all your food and fluids by mouth, you will develop pneumonia from the food getting into your lungs. This is not a "Maybe." This is inevitable. A feeding tube will allow you to get optimal nutrition, fluids and medications, and allow you to continue enjoying those things you can safely swallow as long as possible.

When choking first begins it is time to have a swallow study done. It is unlikely that an early swallow study will indicate that you must quit eating and drinking. Instead, it will show which swallowing muscles are causing the problem and the therapist will give instructions on how to swallow more safely, such as double swallowing, head position, etc. You will be given various textures of food to chew and swallow. The food is mixed with tasteless white goop so that as you swallow, all the stages of swallowing can be seen on X-ray. A therapist can then see how bad your swallow is and recommend ways to make eating safer.

People with even minimal swallowing problems should never eat when home alone. Alcoholic drinks increase the risk of choking and should be enjoyed after a meal, not before. Reduce distractions at meal times. Talking while eating may be sociable, but minimize your part in it.

There aren't any specific rules about what foods to eat, just suggestions. It really is a matter of trial and error to see what foods go down easily. Most people find anything that is crumbly or has dry crumbs, such as chips, cake, cookies, dry toast, corn bread, or hard or crusty bread, etc. cause coughing. But dunking these in liquids, or buttering them, or mixing with ice cream may get them to a safe level for swallowing. Don't try taking a drink after putting these foods in your mouth to soften them. The drink can get to the back of the mouth quickly and trigger a swallow before the food is chewed enough to go down. Drown the food first! Any type of sauce or gravy will help.

Foods that need chewing are a problem not only when the muscles for chewing are weak, but also because the chewing tends to trigger a swallow before the food is well chewed. Meats are especially bad for this reason. Small bites are helpful but I personally find tiny mouthfuls to be unsatisfying! Grinding the meats with a baby food grinder or a blender can help especially with gravy added. For myself, I find hamburger to be a problem. I just can't chew it down enough to get rid of the little pieces that stick in my throat like crumbs!

Raw vegetables are usually the first thing to be taken off the menu, and even steamed or boiled vegetables have to be overcooked to the point where a chef would be mortified to serve them. Casseroles go down well if moist and are a good place to add vegetables and meats. Pasta or rice is easier if it is overcooked too and served with a sauce. Melted cheese and soft breads have to be taken in smaller bites. Even a person without swallowing problems can choke if a mouthful of those get stuck on the way down.

Apples, like vegetables, have to be cooked, but applesauce is to keep on hand. Maybe it is just me, but I find grapes and watermelon to be tricky. One bite and the juice is triggering a swallow before I am ready!

Of course anything that is blenderized or even liquefied is easier to swallow. Smoothies are great but may be difficult to suck through a straw and are better if thicker and using a spoon. No raw eggs added! While balanced nutrition is important, ALS requires a lot of calories, especially

when breathing becomes even a minor problem. ALS patients seem to do better if they can maintain their weight, so calories are good. Protein is important and protein powders can be added to smoothies. Egg custard goes down without chewing and is a good source of protein. A lot of calories can be added with milk and ice cream, but that can cause thick phlegm that is hard to cough up.

Although our swallowing problems are strictly a muscle problem, triggering the sensory system can improve swallowing by stimulating a stronger reflex by whatever muscles are still working although weak. That is especially true for liquids. Liquids are the hardest to swallow safely! They go down so quickly that the weakened swallowing process isn't ready for it. Hot or cold foods and drinks are better than room temperature. A hot or cold drink, especially one with a strong flavor goes down best. Thicker liquids such as orange juice or using a tasteless thickener such as Thick-It will help. The body needs plain water too. It is absorbed without having to go through the entire digestive process to filter out flavorings and doesn't add to the work of the liver and kidneys. A feeding tube makes getting enough water easier.

## *Swollen Feet*

*Note: Although the information here is useful for anyone with swollen feet, it is intended for people with an ongoing problem with swelling of feet and legs due to being unable to walk. If this is not your situation, please consult your doctor to determine the cause and treatment of your swelling. If there is swelling or puffiness of your fingers or around your eyes, see your doctor promptly.*

### *The Cause of the Swelling*

The heart pumps blood through the arteries under high pressure. As the arteries branch out into smaller arteries and then into tiny capillaries, pressure decreases. Oxygen is removed from the blood in the capillaries and then the "used" blood flows into veins for the trip back to the lungs for another load of oxygen. Unfortunately, the pressure generated by the heartbeat has been lost by then and the blood relies on simple back pressure to move back up to the heart. This is aided by muscle activity. Ordinary muscle movement squeezes the veins and pushes the blood along. The veins have little one-way valves all along the way that keep blood from draining backward as it is pushed upwards.

When muscle movement is lost, it becomes much harder to get the blood back up from the legs. It pools in the veins and causes them to get distended. Water seeps from distended veins out into the surrounding tissue and your legs and feet swell (edema). With repeated episodes of swelling, the little veins become damaged and leaky so that water seeps into the tissues even more easily. At the same time, the valves are collapsing under the heavy weight of all that blood that is pooled on top of them. That damage to the valves is **permanent**. Without the valves, the blood pools in the feet even worse than before and remaining valves are under even more pressure and more likely to fail. The circulation to the skin will be affected in time. The skin of the ankles and lower legs will be discolored (bronzing) and the skin fragile. Open sores called stasis ulcers develop. Because the blood flow to skin is poor, these ulcers are very difficult to heal.

### *Treatments*

There is no pill, procedure, or surgery that can prevent leg swelling when walking is limited. It has to be treated by the three words we hate to hear: "Life Style Changes". Minimizing swelling is a do it yourself project!

Doctors aren't very good about helping with swelling. They will offer prescriptions for TED hose (somewhat helpful) and diuretics ("water pills" which should be used as a last resort). And the first thing they will say is to put your legs up to minimize the swelling but they don't tell you how to do that effectively!

Don't be fooled by an adjustable bed or hospital bed. The gizmo that lifts your legs may only lift your knees. Your feet may be left hanging down on the far side of your knees. That is actually worse for circulation to them than lying flat. Put pillows under the foot of the mattress to get the feet back up to the level of your heart.

Look at is the chairs you sit in. A recliner may seem like the ideal way to keep your feet up and swelling down but it is **NOT!** There are two big problems with most recliners. First, the foot rest

section is made in such a way that all the weight of your legs rests on the calves. That is really bad for circulation. Second, putting your feet up -- even way, way up -- without "unfolding" at the hips is minimally helpful as that bend interferes with the already difficult job of moving blood upward to your heart. Lift chairs are wonderful and most of them are recliners, but if you spend most of your time in a recliner, I strongly recommend that you bring the foot rest up only when you lower the back rest. Rather than spending all your time sitting up with the foot rest up, you will probably have better results if you leave the footrest down but take several breaks during the day to recline as flat as possible with the footrests up as far as possible. This self-discipline is so easy to advise but such a nuisance to stick with!

What you need is a chair that can recline fully to what is called Trendelenberg position, feet slightly higher upper body. I found some at [SpinLife](#), a website that specializes in equipment for wheelchair users, but has a large selection of recliners. This page has a comparison chart that shows which chairs have Trendelenberg. These chairs are all recliner/lift chairs and all the pictures show them in a lift position. None show the chair reclined so I can't tell if any have the type of footrest that supports the entire leg, not just propping up the calf. There is a reference to "full chase pad" on some models and that may differentiate the better type of leg rest. If I were buying, I would check that out first. I would also find out about returns if the chair doesn't fit you!

All too often I see PALS and other people in wheelchairs whose foot drop has been allowed to progress to the point where their feet cannot rest flat on the footrest. This guarantees that the feet and lower legs will swell badly and the valves will collapse like dominoes! Don't let this happen! As soon as foot drop droops its ugly head, start using a footrest or positioning boots in bed whenever you are on your back. A footrest can be as simple as a plywood or Plexiglas between the mattress and foot board and pillows in front of it to keep the ankles at a normal angle. While up during the day, wear your AFOs (Ankle Foot Orthotics. These below the knee braces the keep the foot at a right angle to the ankle to prevent tripping.).

Once you quit walking you don't need the AFOs but you do need to keep your feet flat on your footrests. People complain that the footrests are hard and cold. Slippers solve that but aren't as good as wearing shoes for correct positioning of your feet. All too often PALS quit wearing shoes because their toes curl under when they try to get them on. Unless your spasticity is bad, here is how to keep wearing shoes.

- Buy lace up shoes a full size larger than your normal and extra wide (W) or WW wide.
- Remove the lining of the shoes if it is spongy or soft. The inside has to allow your foot to slide in easily.
- Find socks that are thin and smooth for easy sliding. Compression hose work very well. (Don't buy the kind with open toes. They are for hospital use to check for circulation after leg surgery or a cast is first put on.)
- When putting the shoes on, pull the tongue forward to loosen the laces as wide as possible.
- The toes will still want to curl under but unless your spasticity is bad, they should relax after a minute and slide into place. Twisting the toe of the shoe side to side will help get the toes comfortably positioned.

Whether you sit in a regular chair, recliner, or a wheel chair, it must be properly fitted to you. You need to make sure that your leg to floor/foot rest distance is short enough that there is minimal pressure at the back of the lower thigh and knee. Having your feet "dangle" is a sure-fire way to cause swelling! Put a box/platform under your feet (an old hard side suitcase worked great for me -- lightweight and had a handle) or raise your wheelchair foot rest an inch or so. The objective is to

make certain there is minimal pressure on the back of your knees/thighs. If you add a ROHO or other cushion you need to adjust your platform/foot rest upward to make up for the height of the cushion. A note of caution: If your feet are too high, your weight will be shifted back on your tail bone and cause a pressure problem there. It is a balancing act to find the happy medium for footrest height!

Standard power wheelchair leg lifts are fine for adjusting your legs while sitting up, but when you lie back in your chair and raise the footrests, the footrests are suddenly too short! Your knees have to bend or you need a big pillow to get your heels above the footrests. Very inconvenient and hard to get comfortable! The solution is to order "articulating" leg rests. These lengthen as they lift so that your legs aren't scrunched even with the legs all the way up. Comfortable for elevating your feet to reduce swelling or just catching a nap!

The best treatment for leg swelling that I have found is something that I discovered entirely by accident: **More time in bed.** When my husband was working, I spent about seven hours in bed at night and then would lie back in my recliner for another two or three hours in the afternoon. Even with that, my legs were swollen by noon, miserably uncomfortable by evening and absolutely painful by bedtime. When my husband retired, I was able to go to bed at the usual time, listen to books on tape for an hour or two, and then sleep late in the morning. Instead of spending 10 hours lying with my feet up in two separate sessions, I began spending 10 hours or more in bed all at one stretch. Within a matter of days after starting this routine, I noticed that the swelling was minimal. Now I don't even have to lie down in the afternoon in order to be comfortable in the evening! I don't know if this is due to spending more time lying down at one stretch, spending all my lying down time in a bed rather than a recliner, getting more sleep, or some combination of the three. All I know is that in this has made an incredible difference for me. Not only has it made my problems with swelling minimal, I feel better in general.

Another thing that helps is **muscle activity**. Granny's old rocking chair served a real purpose beside putting babies to sleep! I find that the swelling is minimized on days when I am most active. (Interpret that as days when I am frequently hauled in and out of my chair and forced to stagger a few steps, whining all the way!) I guess I have some muscles left in my legs, even though I sure can't feel 'em! Even passive range of motion exercises help.

Keep cool. A few minutes of being too warm, toasting my feet by the fire, or just sitting in the summer sun is all it takes to turn my feet into balloons. (Blood vessels dilate when we are warm.) Simply keeping my legs in the shade makes a difference, but I have also been known to pour cold water over my feet on hot days when I need to be outside. Wet socks and tennis shoes are still more comfortable than that miserable burning sensation of swollen feet!

Sometimes I also have problems with a burning sensation in my feet in bed at night. It doesn't start until my feet began to warm up. It can get really bad in the middle of the night if I have the electric blanket on and my feet get really warm. That is a real nuisance because the rest of my body gets really chilled and I can't move at all if I pile on extra blankets. So, in cold weather I end up sleeping with the electric blanket on, but my feet sticking out!

For some people, this burning pain becomes severe and doesn't seem to be relieved by getting the swelling down. This might be the end result of long term or severe swelling. Some people find that aspirin (not Tylenol) helps. Do not take aspirin if you are on anticoagulants (medications to thin the blood). If burning pain is felt when swelling has not been a problem, discuss it with your neurologist.

**Limiting salt intake** used to be high on the list of things to do to minimize swelling, and your doctor may suggest it, but the need for that is questioned these days. I guess it is enough to say don't over-indulge with salty foods.

Hospitals often use **devices to improve blood flow** to the feet of patients who are going to be stuck in bed for a while in order to reduce the risk of blood clots. TED (elastic or compression) stockings are by far the most common. By simply squeezing the legs and feet a little, they help keep the veins from getting distended. You can ask your doctor for a prescription for these stockings, but unless you have strong hands and arms, you will need help putting them on.

Hospitals also use Sequential Compression Devices that inflate and deflate to help pump the blood along. Originally used for hospitalized people at risk of blood clots, they are now available for home use to improve blood flow. This is very effective in reducing swelling as well as the risk of blood clots in people who are not able to walk. They have some type of leg sleeves or boots or wraps that are connected to a pump that causes compression in a sequential upward direction to improve blood flow. With help from your doctor you may be able to get your insurance to cover the cost of this equipment. It is not complicated to use, but you must be very careful to make sure that it is not rubbing anywhere and causing breakdown of the skin. SCDs can be used while up in a chair which makes using them convenient.



Another option is a leg massage device. They are not medical devices and are probably less effective for swelling than compression devices but may be very helpful. They don't squeeze the blood upward, just massage the feet and legs. One big advantage is that they don't require any type of boot or wrap, just placing your legs in the massager which is easier. Massagers are generally used while up in a chair.



If you complain about swollen ankles and feet to your doctor, odds are he will whip out the old prescription pad and put you on **diuretics**. I have real reservations about this because many of us are borderline dehydrated half the time anyway. (Another contributing factor for the development of blood clots.) It gets hard to reach a drink, or hard to swallow, or it is simply too hard to get to the bathroom so we don't drink as much as we should. Diuretics cause your kidneys to remove more water from your blood stream. The thicker blood is then able to sponge up more water on its travels through the body so it does reduce the edema. It does nothing about the cause of the edema, poor blood flow, however. Using diuretics for swollen legs is kind of like taking a diuretic to lose weight -- sure it "works", but it doesn't really solve the problem.

I certainly won't say diuretics should never be used. If nothing else works well enough to keep the swelling under control, diuretics need to be used because the swelling further damages the veins and valves and the situation just gets worse. But all the things described above should be implemented first before diuretics are considered.



## ***Prevention and Treatment of Blood Clots***

Your doctor may recommend that you take a blood thinner after you begin using a wheelchair full time because you are at high risk of developing blood clots, especially in your legs when walking is minimal. If a blood clot forms, the danger is that pieces of the clot will break off and sail through the blood stream looking for places to get stuck and cause major damage. Clots in the lungs (pulmonary emboli) cause severe chest pain and breathing difficulty. Clots in the brain cause strokes. The recommendation may be for an Aspirin daily, or if you have had a clot, he will want you to take Coumadin (warfarin). Many people are reluctant to take any medications and the idea of any blood thinner, much less the one that is the lethal ingredient in rat poison, sounds pretty scary! Newer anticoagulants that do not require regular checks of your clotting time are available (and heavily advertised!). These can be valuable if you can't leave home or don't live near a hospital or clinic where the test can be done.

1. The choice of which medication to use is dependent on several factors:
2. Coumadin (warfarin) is very inexpensive, as low as \$50 per year. The newer drugs are far more expensive at around \$3000 per year.
3. Coumadin requires monthly checks of clotting levels, the newer meds do not.
4. Coumadin's low price and once a day dose make it more likely that the patient will comply than the high price and twice a day doses of the new meds.
5. Patients may be more compliant with the newer meds if Coumadin testing is not easily available to them.
6. The level of anticoagulation of the newer meds cannot be determined by any blood checks so insufficient anticoagulation can't be detected.
7. The effects of the newer meds can not be quickly reversed in the event of a hemorrhagic stroke, emergency surgery, or trauma causing severe bleeding. Coumadin is quickly reversed with an injection of vitamin K.

### **Clot Watch!**

Any pain in the leg should be taken seriously. If the leg or foot swells, the area is tender or painful, hot to the touch, red, or having your toes pushed upward makes it hurt, you need to see a doctor right away. Don't massage it or wrap it or anything else, just get your resistant, obstinate arse to a doctor or E.R. An ultrasound of the leg needs to be done and a typical doctor's office doesn't have the ultrasound equipment. You will probably end up having it done at a hospital anyway, so your doctor will likely send you there rather than the office. If it is a clot, you will promptly find yourself in the hospital with a Heparin IV. Yeah, argue and groan all you want, but this is necessary.

Coumadin takes several days to reach an anticoagulating level and during that time your clot will get bigger and bigger. Heparin IV works immediately. You will be on Heparin for several days. Heparin does not break down the clot, just keeps it from enlarging. Your body will gradually break down the clot. While you are on Heparin, Coumadin will be started. The two medicines don't work on the same step of the clotting process so you will need to be on both until your blood tests show that the Coumadin has kicked in and is at the right level. Then, and only then, will the Heparin be stopped and you can go home.



8. New meds are not affected by diet. Coumadin doses can be adjusted to any fairly consistent diet. Only large changes, especially in foods high in vitamin K are likely to be a problem.
9. Generally Coumadin is used for financial reasons if it is likely that the patient can and will follow through with regular testing. The newer meds are used if that is not likely or if the patient is at high risk of severe and potentially fatal clots.

It is also possible to use subcutaneous injections (just into a pinch of skin and fat, not into the muscle) of Lovnox instead of Heparin. Lovnox is usually given on your stomach, twice a day. It stings pretty bad and leaves bruises, but it can be done at home with the help of Home Nursing Service. Your doctor won't like this option though, and getting the nursing Service lined up quickly may be a problem, but being in the hospital is not at all a good thing for an ALS patient! A visiting nurse can come to the house to give the injections or teach you to give them. These injections are not into muscle, just under the skin. The medication is in a pre-filled syringe so the dose is always accurate. The injections sting for a few minutes and leave bruises from the concentration of the anticoagulant. Putting pressure on the injection site for a couple of minutes may reduce the bruising. Not fun at all but beats the heck out of being in the hospital when you are helpless. I have done it both ways and being at home is my preference.

Heparin can also be given subcutaneously in small daily doses. The advantage of using heparin is that it provides rapid but short term anticoagulation which makes it ideal in cases of severe trauma or surgery where late bleeding is a possibility.

The bottom line is that a blood clot is far more dangerous than carefully regulated anticoagulated blood running through your blood stream.

I have been on Coumadin for about 6 years after having a blood clot in my leg. I have had absolutely no problems with it, not even a tendency to bruise very easily. The regular lab work to check my clotting time was a problem at first because drawing blood from me is difficult. Also, it required waiting for the doctor to get the test results and call me to let me know if the dosage should be changed and when I should be checked again. The only good part was that I didn't have to have an appointment to have the blood drawn.

Then Coumadin clinics began opening. They use a simple finger stick to get a drop of blood, run it through a little meter, and have your results in seconds. The nurse then decides if you may need a dosage adjustment. If so, she will talk to the pharmacist who is part of the clinic and he/she will adjust the dosage. The nurse schedules your next appointment and you are done.

Frequency of appointments varies. If your clotting time changes significantly they may want to recheck in a week or two. If it is where it should be, the next check won't be for a month. And if it remains stable it may be 5 or 6 weeks.

What you eat does affect your clotting time. Foods high in vitamin K, such as green vegetables, make you clot faster. *That doesn't mean you can't have those foods!* It just means not to eat a lot more or less of them than you usually do in the course of a week. Your Coumadin dose is adjusted to your routine diet, whatever that maybe. You don't have to change what you normally eat, just save yourself the hassle of frequent checks and dosage adjustments by keeping the amount of greens fairly consistent week to week. Most people end up having their Coumadin dose increased a bit in the summer when fresh veggies are eaten more often.

Lots of medications affect your levels. Tylenol is fine but anything with Ibuprofen will affect it. Again, that is not a problem if you take Ibuprofen on a regular schedule (your Coumadin will be adjusted for that) but if a new ache or pain has you taking it, expect changes in your clotting time.

- Antibiotics really screw up your levels. If you have to begin antibiotics, call the Coumadin Clinic. They can adjust your Coumadin to minimize the change and schedule you for a recheck sooner to avoid big swings in your levels.
- A lot of other meds, prescription and over the counter, affect Coumadin so you will be asked about any changes in your meds at each check.
- Infections, stress, and just about anything can also affect coagulation level so regular checks are very important.

As far as injuries while on Coumadin, cuts and scrapes will stop bleeding, they will just take longer. Big cuts will require stitches anyway, and an ER visit needed. Just as with any fall or injury, any internal pain or sign of a brain injury (loss of consciousness, even momentary, headache, dizziness, confusion, drowsiness, vision or speech problems) requires attention. And, no, you won't automatically die from a head injury just because you are on Coumadin

If you should require surgery while on Coumadin, the surgeon will stop the Coumadin a couple of days before surgery. In an emergency, he can use Vitamin K to counteract the Coumadin. Wearing a medic alert bracelet is probably a good idea. If you need dental work beyond cleanings or fillings, your dentist may want you off Coumadin, but generally that isn't necessary even if you are having a tooth pulled. Trach and feeding tube changes don't normally require you to stop the Coumadin either.

As you would with any medicine, keep your TicTac size Coumadin out of reach of children. A shot of Vitamin K is the antidote for Coumadin but it is still a poison in large doses.

So, bottom line is that being on Coumadin is only a minor hassle if you are portable enough and near enough to get to a Coumadin Clinic regularly. And going on Coumadin before you develop the almost inevitable blood clot beats the heck out of a week in the hospital or belly shots!

## *The Right Calcium Supplement in ALS*

As improved care options and, hopefully, treatment advances extend the life expectancy of ALS patients, more of us will need to address long term problems such as osteoporosis. As we enter middle age, we are constantly reminded of the need for calcium to prevent osteoporosis. Osteoporosis occurs in immobilized people as well as in older people. The ALS patient needs to understand the processes at work because the standard treatments for age-related osteoporosis are not helpful and potentially harmful for the disabled.

### *Bones and Calcium*

Bone is living tissue that is continually being broken down and replaced. Up to age 30, more bone is formed than lost, but after age 30 that changes. Although we continue to replace bone, we have an overall net loss of bone mass. That loss is normally very gradual but is sped up by some factors. It is affected by race, heredity, body type, age, and especially by gender. Women have more bone loss than men because of hormonal changes that occur with menopause. Diet and smoking have an effect on bone loss, and lack of exposure to sunlight can prevent absorption of calcium from the food we eat. Another factor is decreased exercise. That is a small but important factor for the typical osteoporosis patient, but is the major factor for the long term ALS patient.

The trigger that makes bone absorb calcium and keep itself strong is stress on the bone. Exercise, especially exercise that involves bearing weight on the bone, stimulates the bone to absorb calcium and rebuild itself. When we are immobilized, calcium continues to be slowly lost from the bone in the normal way but is not replaced because the trigger to replace it, exercise, is gone. In disabled people, the problem is not that we lose bone mass faster, it is that we fail to replace it. The result is osteoporosis -- weak, brittle bones that break more easily. That is a real problem for ALS patients as we are prone to falls.

For the immobilized patient there is another problem seldom discussed in the literature on osteoporosis because it is so oriented to the osteoporosis of aging: The calcium being lost from the bones and not being reused to rebuild bones but ends up circulating in the blood. Lab tests of blood calcium will show abnormally high levels. (There may be some cellular problem with the use of calcium that leads or contributes to the loss of motor neurons, but the high calcium levels seen in ALS patients is NOT a cause of ALS but a result of the immobility ALS causes.)

The kidneys will filter out calcium and excrete the excess, but, in time, the kidneys will get clogged up with calcium. Kidney stones will form and all the problems associated with kidney stones can occur. This is not uncommon in the general population and it is a very common problem for spinal cord injured patients. For ALS patients this has rarely been a problem simply because life expectancy is shorter than the time needed to develop kidney stones. Long term ALS patients may face this problem however.

### *Treatment*

Although early diagnosing of osteoporosis is difficult, when a person with ALS shows up in the Emergency room with a fracture from what should have been a "survivable" fall (one not causing broken bones) it is logical to assume that there is a significant amount of osteoporosis present. But what is the proper treatment? This is where the literature leads us (and sometimes our

doctors) astray. Applying standard treatments for osteoporosis to ALS patients may cause worse problems. The standard treatments are:

*Increased dietary intake of calcium.* There are two problems with this treatment for ALS patients. First and most significantly, our problem is not lack of calcium or even our loss of calcium from the bones, but rather the lack of the trigger to move available calcium back into the bones. Increasing intake of calcium only leaves more calcium in the blood. That doesn't help the bones and can cause kidney stones. Second, dairy products are the biggest source of calcium but dairy products tend to thicken mucus and respiratory secretions. ALS patients already have enough with choking and gagging and may need to be restricting their intake of milk and milk products, not increasing it.

*Calcium supplements (oral, intravenous, or injected).* The problem is the same as above. We don't need more calcium, we need the trigger to move it back into the bones to replace normal bone loss.

*Sunlight/Vitamin D* Oral calcium requires Vitamin D to be absorbed but more importantly for the ALS patient, Vitamin D, especially D3, helps move calcium into the bones, making it is a valuable treatment for the ALS patient. The best source of Vitamin D is our own skin. Sunshine causes the skin to manufacture a substance that our liver turns into Vitamin D. Just 10-15 minutes of sun 2-3 times a week is all that is required so there is no need to risk skin cancer!

Although dietary intake of Vitamin D is normally not especially significant, people who can't get sunshine can increase intake with foods. For decades, our milk has been routinely fortified with it to prevent rickets, but milk may need to be avoided by ALS patients for the reason discussed earlier. Some breakfast cereals are fortified with Vit. D (read the label), and butter and eggs, fatty fish (such as herring, mackerel and salmon) are sources. As a last resort, Vit. D supplements are available. Don't take more than 400 IU of vitamin D a day unless prescribed by your doctor. Excess is not excreted so you can overdose. Vitamin D toxicity can lead to nausea, weight loss, irritability, and formation of calcium deposits in your lungs, kidneys and soft tissues.

*Estrogen* Estrogen is the most common drug prescribed to preserve bone mass. Estrogen has been proven to prevent menopause-related bone loss but it is apparently not useful in other cases of osteoporosis.

*Bisphosphonates* Bisphosphonates such as Fosamax and Boniva are now being prescribed. These are non-hormonal agents that prevent bone from releasing calcium. This treatment works to alleviate the inability to replace calcium lost from the bone and is logically a better choice for the ALS patient than treatments that only add calcium to the blood stream.

In summary, ALS patients are prone to osteoporosis and the resultant brittle bones, but it is related to their inability to exercise as much or more than the typical factors of aging. Blood calcium levels will already be high, and additional calcium won't help and may even be harmful long term. ALS patients can best combat osteoporosis by getting what exercise they can (and for us, simply being held up in a standing position is the weight bearing exercise needed), and by making certain we have an adequate amount of Vitamin D by getting a little sunshine. If further treatment is needed, discuss with your doctor whether his proposed treatment that will get the calcium into the bones, not just into the bloodstream.

# *Toilet Troubles*

## *Constipation in ALS*

Although we joke about constipation, it is a miserable experience and should never be taken lightly in the ALS patient. Loss of appetite from frequent constipation leads to weight loss, weakness, and dehydration. Constipation can progress to blockage in the intestines and nausea, vomiting, and abdominal distension. (Vomiting is very dangerous for a person who cannot turn over when lying on his back because it causes choking.) One early sign of blockage is often overlooked. Repeated small loose or liquid stools may be ignored or thought to be sufficient when they are actually the result of a large amount of hard stool blocking the bowel with only liquid stool being able to pass around it. The blockage can become so severe as to require hospitalization and possibly even surgery to correct.

### *What is constipation?*

That may sound like a foolish question, but many people think of constipation as having infrequent, dry, hard bowel movements. It is actually defined simply as having stools that are hard to pass. Many people have only a couple of bowel movements a week, but if they do so without straining, they are not constipated.

Normally food is liquefied in the stomach by digestive juices and moves through the small intestine in liquid form. Nutrients are absorbed in the small intestine. Waves of muscle contraction called peristalsis move the remainder along into the large intestine. In the large intestine, water is reabsorbed from the left over waste product, leaving just fecal material (stool) which is moved along and passed out of the body in a bowel movement.

Anything that changes the speed with which foods move through the large intestine interferes with the re-absorption of water and causes problems. Rapid passage causes diarrhea, slowed passage allows too much water to be reabsorbed, leaving hard, dry stool that doesn't move easily through the bowel.

Common causes in ALS patients include:

- ◆ Many medications affect bowel function. Prescription pain medications are especially constipating.
- ◆ Certain foods, a poor diet or changes in diet.
- ◆ Constipation is very common in anyone with poor mobility because lack of activity and exercise slow bowel motility.
- ◆ Long delays in getting to the bathroom further complicate the problem by keeping the stool in the large intestine longer where it becomes drier and harder.
- ◆ There is some evidence that ALS can affect the autonomic nervous system as well as skeletal muscle and slow the entire digestive process.
- ◆ In ALS swallowing problems make getting a good diet and sufficient fluids and fiber difficult and the problem gets worse.

Breathing problems make it difficult to take a deep breath and bear down, something we don't even realize is important in having a bowel movement until we cannot do it.

Because so many things contribute to constipation in the ALS patient, the solution may change over time.

## **Diet**

The first -- and best -- way to approach constipation is by improving your diet.

- ◆ Drink lots of fluids.
- ◆ Eat lots of high fiber foods. (Check with your doctor if you have other digestive or bowel problems or are on a special diet.) There are many high fiber cereals available and granola bars are convenient and easy to handle when feeding yourself begins to be difficult. Raw fruits and vegetables are also easy to eat sources of fiber if swallowing is not a problem.
- ◆ If you are using tube feeding, fiber is added to most tube feedings formulas. Check the label to see if your brand has added fiber.
- ◆ When diet alone isn't quite enough, try the old remedy of prunes or prune juice for occasional constipation. It really does work!

## **Laxatives**

### *Fiber*

- ◆ Fiber laxatives supply the fiber necessary to add bulk which holds water and makes it easier to move the stool through the bowels.
- ◆ Today's over-processed foods are low in fiber to begin with and when swallowing problems begin there is usually even less fiber in the diet.
- ◆ Fiber laxatives are very slow acting and are taken daily to prevent constipation rather than for relief of existing constipation.
- ◆ Generally the first laxative recommended for frequent constipation, fiber laxatives are also ideal for long-term use because the fiber is not absorbed.
- ◆ Two well-known brands are Metamucil and Citrucel. Available without a prescription, some use natural fiber (agar, psyllium, kelp and plant gum.) Others are synthetic cellulose (methylcellulose). Natural and synthetic bulk-forming laxatives act similarly.
- ◆ Fiber laxatives are available as a powder (which is mixed with water or juice and generally needs to be swallowed fairly quickly before it thickens to a goo, though newer brands without that problem are available.), a tablet, or a wafer.
- ◆ It is possible to be allergic or sensitive to flavorings or other additives. Some brands may also contain enough sugar as to cause problems for diabetics.
- ◆ For the ALS patient there are two concerns with this type of laxative;
  - It is essential that fluid intake be very good. 8 ounces of fluid must be taken immediately with each dose and more throughout the day is needed for safe, effective use. Taking fiber laxatives without enough fluid can cause intestinal blockage.
  - They are not to be used when swallowing problems begin. Failure to drink enough water to wash down the fiber might allow it to begin to swell in the esophagus and this requires immediate medical attention. Fiber laxatives can safely be given through a feeding tube, but the fiber needs to be promptly followed by flushing the tube with water to prevent clogging.

### *Stool Softeners*

- ◆ Stool softeners, also called emollient laxatives, also keep the water content of the stool higher which keeps it softer and allow it to move more easily through the bowels.
- ◆ Stool softeners are often ideal for ALS patients. Not only do they help keep the stool soft when fiber and fluid intake is difficult, but they also are very helpful when breathing problems make it difficult to bear down and push.
- ◆ They do not cause frequent bowel movements, cramping or urgency but greatly reduce the amount of straining needed to have a bowel movement.
- ◆ Stool softeners are taken daily as a preventive measure rather than to force a bowel movement on a certain day.
- ◆ Stool softeners are available in pill or liquid form.
- ◆ Colace is the most commonly prescribed stool softener, but there are many non-prescription brands of the active ingredient, docusate, available, such as Surfak. Liquid docusate is also available without a prescription but the pharmacist will probably have to special order it for you as it is seldom stocked by drug stores. (Note: liquid docusate needs to be diluted in juice for drinking or it burns all the way down!!!!)

### *Hyperosmotics*

Hyperosmotic laxatives draw water into the bowel from surrounding body tissues, softening the stool. There are three types of hyperosmolar laxatives.

- ◆ The saline type is the most well-known -- and disliked! Saline laxatives are harsh, fast acting, and total in effect. They are primarily used to completely clear the bowel in preparation for surgery or bowel exams.
- ◆ The polymer type is a large molecule that causes water to be retained in the stool to soften it and increase the number of bowel movements. It is not used long term.
- ◆ Of the three types of hyperosmotic laxatives only one, lactulose, is useful for preventing constipation. Its action is so much less rapid and harsh than the saline that it is often used for long-term treatment of chronic constipation. Because it has sugar-like properties it may not be suitable for diabetics. Lactulose is available only by prescription.

### *Lubricant Laxatives*

- ◆ Lubricants use mineral oil to coat the stool for easier passage. Mineral oil should not be taken by patients with even the slightest swallowing problem. Aspiration of oil into the lungs causes chemical pneumonia.

### *Stimulant Laxatives*

- ◆ The majority of non-prescription laxatives are stimulants and contain senna, castor oil, cascara, aloe, bisacodyl, or combinations. These laxatives are often marketed as being safe, "natural" remedies because the active ingredients come from plants. That makes them natural but, like many other plants, they are basically poisonous. That is why the body finds them irritating and reacts so quickly to get rid of them.
- ◆ Stimulant Laxatives increase the muscle contractions (peristalsis) of the bowel which moves the stool along. Most are intended to be fairly gentle and result in a bowel movement within 6 to 12 hours, but even these can cause cramping. If constipation is

already making you uncomfortable, stimulant suppositories will provide relief within an hour but are likely to cause cramping.

- ◆ Stimulant laxatives are not for continuous or long term use! Even in ALS, they should be reserved for occasional use until other methods fail. Frequent use of stimulant laxatives can actually aggravate constipation because the bowels become dependent on them for the stimulation for even normal peristalsis. These laxatives work by irritating intestinal nerve endings, which in turn stimulates muscle contractions that move the irritant through the gut and out of the body. After a while, the nerve endings no longer respond to this amount of stimulation and larger doses are needed. For long term ALS patients, after years of frequent use, the nerves of the colon slowly disappear, the colon muscles wither, and the colon becomes dilated and unresponsive to laxatives.

### ***Enemas***

Enemas are sometimes necessary when diet, fiber, and laxatives fail, but are generally the last resort. They are usually used to remove impacted stool higher up in the large intestine and may be the last step before surgery. Regular use will, over time, dilate the rectum and lower bowel. This reduces the ability to have normal bowel movements without an enema. "Mini" enemas are now available. They contain stimulant laxatives to stimulate bowel movements. They contain less fluid so they don't dilate the rectum and lower bowel as much so would be preferable to a standard enema such as a Fleet's enema, or the old fashioned soap suds enemas.



## ***Women's Lib: Urinary Options***

Women just don't have the plumbing to make urinating easy when transferring to the toilet gets hard. There are several possible solutions, none of them perfect, but every one is better than not drinking enough to delay having to go. That leads to chronic dehydration and bladder infections, constipation, and lung problems. The first step in solving the problem is admitting that you can no longer manage urinating without assistance. As is often the case with ALS, the result of insisting on doing things by yourself only makes things harder, riskier, and more stressful than they need to be.

Having help with something like urinating that has always been a private activity is a big step for most women. It is a big step for family helpers too. I still laugh when I remember one of the first times my husband had to wipe my bottom. The back of his hand over reached and dipped into the toilet. He jerked it back as though he had been scalded and jumped around the room moaning about having touched pee. His reaction was so much like a hysterical four-year old. I really expected him to regress to calling it pee-pee! That reaction is long gone and he deals calmly with all toilet details.

Urinary Catheters, sometimes called Foley catheters, take the worry out of needing to urinate so are often used by people who are home alone for longer than they can "hold it". The most commonly mentioned drawback to a catheter is the risk of urinary infections. Some people seem to be plagued with repeated infections, others never or seldom get them.

I am resistant to the idea of using a catheter for other reasons. Having had one while hospitalized, I know how incredibly painful it is to have the tubing get hung up and pulled during a transfer. It isn't a pain that ends when the pulling stops either. I found that sitting on the tubing was not comfortable for my private parts either.

Some women opt for a suprapubic catheter instead. This is a catheter that is surgically placed through the lower abdomen. The biggest drawback seems to be bladder infections, but again, some people don't get them. Some people have leaking around the tube. Many doctors are resistant to the request for a suprapubic because they don't see it as need, just a convenience not worth the potential problems. The convenience is considerable. As with the regular urinary catheter, it can be attached to a large collection bag or a smaller, more easily concealed bag. It can also be clamped and opened and drained when bladder fullness is felt. There is the added work and expense of keeping the bags clean and odor free, so there are several important pros and cons to a suprapubic catheter.

Before resorting to a catheter, I strongly encourage all women who aren't bed bound to try a female urinal. They have a curved cup that fits up against you to prevent leaks. They only work if you can be scooted /slouched forward in your chair so that you are slightly over the edge in front. For anyone with weak arms or hands, using a female urinal will require assistance. You can't wear panties and will have to be wearing a skirt or open bottom slacks to use it. These adaptations have been 100% worth it to me because I never need to worry about a full bladder no matter where I am. See ***Adaptive Slacks*** for a little more info on urinals and adapted slacks.



When buying a female urinal, find one with the longer section of the cup on the bottom. If it is at the top, it is only only useful if you can stand to urinate. "Spill proof" means that after use it will not spill if it tips. It doesn't mean leak proof while in use if it isn't snug against you! Spill proof is ideal for traveling or to keep in your vehicle. Spill proof urinals have a removable cup that can be

positioned for standing or sitting. Unfortunately, pictures show them in the stand to urinate position which is confusing. I am not even sure why anyone who can stand needs a urinal!

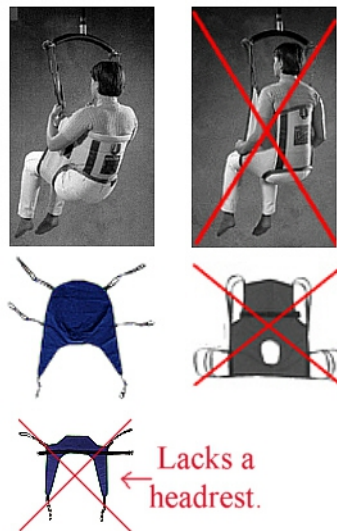
This female urinal has worked very well for me. I have had few leaks since the first time I used it. We experimented and found that by reclining my wheelchair a bit, using the towel that I sit on to scoot me forward in my chair, and then placing my feet to the outer edges of the footrests, it is easy for a caregiver to position and doesn't leak. I wear open bottom slacks all the time but this would work with skirts as well. The urinal has a one-way valve in it so it can't be spilled after it is used. It comes apart easily for emptying and rinsing or washing. I bought it for use on a trip and now I keep one in the van so now my time away from home isn't dictated by my bladder. It is easy to use privately in the van and is definitely on my must have list. My husband prefers it to transferring me to the toilet so we use it at home as well. The urinal with the anti-spill section and the curved cup attached is rather large. It takes a few tries to learn how best to position yourself, and unless you have good arm strength, you will need assistance.

## *How to Use a Lift for Toileting*

One of the most frustrating problems presented by ALS is using the toilet. For men, finding someone strong enough to lift them is a problem. For females, this is a hassle faced several times a day. The most frequently recommended solution is a lift system such as a Hoyer lift or ceiling track lift. Either type eliminates the need to lift the patient and that is a big help, but information on this equipment focuses on lifting and transfer ease. Nobody mentions getting your pants down! No one tells you that in order to use a sling lift for toileting, you have to give up wearing slacks and underwear! Well, I refuse to sit around bare-assed under a drafty skirt or lap blanket waiting for a call of nature! With a bit of experimenting, I've found a way to dress normally and still use my lift.

The brand of lift you get doesn't make as much difference as the type of sling you use with it. Despite what lift manufactures want you to believe, most slings will work just fine on most brands of lifts. Refer to Lifts for information on the types of lifts available and suggestions.

The first thing you need is the right kind of sling, one called a "split leg", "hygiene" or "U-sling", NOT a "transfer" or "commode" sling. Commode slings simply have a hole under your bottom. Hygiene/toileting slings are open underneath your bottom and all the way up to your waist in the back. The key feature I wanted in a sling was that it be easy to put on and take off while I was in my wheelchair. I did not want to have to sit on it all day. Besides being a red flag for the fashion police if I wanted to go out, it could also contribute to pressure sores. These pictures show the difference between a usable toileting sling which can easily put on and removed while seated in a wheelchair, and a commode sling which cannot.



## *Dress for Success*

A feature I needed was to be able to get my slacks down and back up without a big hassle. Some manufacturers of toileting/hygiene slings say your caregiver may be able to get your slacks down and back up again while you are in the sling but none guarantee it. We found that with some heavy tugging we could get them down, but getting them back up was a major effort and soon abandoned. That led me to try adaptive clothing.

First, I tried slacks that have zippers on both sides. When unzipped, the backside of the slacks drops down like a trap door and can be pulled forward out of the way before you are lowered down

onto the toilet. Lifting the flap back up before sitting requires a couple seconds of standing with only one armed support. Getting the zippers re-zipped once back in the chair required dexterity and hand strength and frustrated my helpers. Side zip pants would still be the best choice if you ever have to stand up in public, however.

A much better solution is slacks with an open back. They are far easier to get on in bed or in a wheelchair. As weird as they sound, they look like ordinary slacks when you are sitting in them. Of course, you can't wear underwear with them. I just put a hand towel on the wheelchair seat instead. They can be put on while you are lying in bed or sitting in your chair or on the toilet. Just have your helper put your feet in, pull them up to your thighs, lift your knees and pull them up and tuck them under, and snap the waist band in back.

When you transfer to the toilet -- by standing or in a sling -- you don't have to do anything with them. Just transfer to the toilet and go! No removing, pulling, unzipping, unsnapping, un-Velcro-ing needed. There is plenty of open space underneath -- just move the flaps to the outside of the toilet or bedpan. Back to your chair, remove the sling, do a quick fanny check of your sides to make certain there isn't a revealing gap, and you are on your way. A pit stop with the help of a caregiver who is familiar with the process takes less than 10 minutes --even with me on a ventilator now!

### *How to Use a Toileting Sling*

A sling takes practice to get on properly. You really can't slide down through it as you are lifted, but your position can be adjusted to get you sitting fairly upright. If you think of it as being like the seat on a kids swing where your weight is on your upper thighs rather than your butt, the mechanics of positioning it makes more sense. My husband was ready to throw the sling out when it didn't work the first time. I had to insist that we take the time to try different loops on the straps, cross the leg straps, and get the leg straps further up under my thighs. Hope you have a little more patience!

- Place the sling behind the patients back with the lower edge of the sling at the level of the waist band.
- If you wear side-zip slacks, unzip them.
- Bring the leg straps forward and under the leg, lifting each knee to get the straps up high under the thigh. This is the key to a safe, comfortable transfer! If you slide butt down too far through the sling, it is probably because the leg straps need to be moved further up under your thighs.
- Fasten sling safety belt. (Actually, we ditched the belt. With the back rest and leg straps correctly positioned, I don't slide down and can't tip out. I am no daredevil -- I wouldn't use it that way if I didn't feel safe!)
- Bring the lift bar over the patient and lower it. Hook the loops of the straps over the hooks on the bar. Each strap has a series of three loops. By choosing higher or lower loop you can adjust whether you sit upright or lean back. I find it more comfortable (and dignified!) to cross the leg straps in front of me to keep my knees together.
- If the patient is to be moved sideways to the toilet, remove the armrest on that side.
- Recheck to make sure none of the loops have slipped off the hooks and begin lifting. After making sure the patient is not slipping down through the sling (indicating that the straps need to be moved up higher under the thighs), move the patient over the toilet.

- If you wear side-zip slacks, grab the back waist band of the slacks and pull it down and forward while lowering the patient onto the toilet. For women, there is no need to pull down the front of the slacks.
- When the patient is finished, recheck to make sure none of the loops have slipped off the hooks and begin lifting. When the patient is a few inches above the toilet, wiping can be done easily.
- Lower the patient back into the wheelchair. If you use side-zip slacks, grab the back waist band of the slacks and hold it back up in place while lowering the patient back into the wheelchair.
- Remove the sling, zip slacks, etc.

### *Purchasing a Toileting/Hygiene Sling*

Although there are several manufacturers of the right type of sling, you may find they will not sell you the one you want unless you have their same brand of lift. This is no doubt explained as a liability issue, but the truth is that nearly all slings will fit nearly all lifts. The most common incompatibility is that the sling has four or six straps and the bar (sometimes called the carriage) of the lift only has two hooks. As long as the straps of the sling have loops so you can select the length you need, you should be able to be positioned upright or at a comfortable angle even with just two hooks.

The easiest way to buy is to select a brand of sling sold by general medical supply web sites. There you can purchase over the Internet without having to speak to anyone. Just put one in your shopping cart and pay for it.

If you really want a sling sold only by a lift company, be prepared to lie! Read their website and decide which model of their lifts you are going to claim to have. If they ask for the serial number, try "Oh, I don't have it yet! I just know it will be the model 700M. My brother-in-law is supposed to be going to get it but I don't know how soon that will be." (Everyone can relate to the undependable brother-in-law story.) If they want to send a sales rep out to "make certain it is the sling that will best suit your needs", hang up.

Google "hygiene sling" to get started. Invacare and Hoyer are good brands sold at many websites. Prices on the same sling vary widely among suppliers. Once you find the brand and model you like, do a web search for the name and model number for the best price on it.

### *Links to Clothing*

The two main suppliers for open back slacks are Silverts and Buck and Buck.com/. Fabrics and colors are limited. You will find that adapting your own slacks or having it done allows you complete control over fabric, color, fit, and style. Jeans, shorts, men's or women's! My only restriction is to avoid thin fabric that will tear easily. Alteration instructions here; *How to Adapt Your Slacks*

## *Adaptive Slacks for Easy Toileting*

*The option of wearing slacks is yours again!*

After battling with the problem of dressing and using the bathroom when I could no longer stand up long enough to have my slacks pulled up and down, I discovered the incredible convenience of open back adaptive slacks. Yes, they sound embarrassing, but how often do you jump up out of your chair in public? Unless you stand up, no one can tell that you aren't wearing ordinary slacks!

The pros far outweigh the single con of not wearing underwear under them. Underwear is not recommended for people in wheelchairs anyway -- it adds seams that can cause pressure sores. Instead, put a hand towel on your cushion (long side front to back) and sit on that. In addition to being comfortable, absorbent, easy to change and wash each day, a towel can be pulled from behind your back to scoot you up in your chair.

The pros are:

- Easy dressing. No hard tugging required.
- Super easy transfers to the toilet and back again whether you are being lifted to transfer or using a sling and lift. The open bottom means your slacks don't have to be tugged down and then up again. Just lift and sit! The opening keeps the slacks well out of the line of fire and makes wiping easy.
- They even make using a female urinal easy for women. Your caregiver just scoots you down in your chair enough that you can pee over the edge, puts the urinal firmly in place against you, and you can go without the hassle of finding an accessible bathroom. When finished and wiped, your caregiver can scoot you back up using the towel you are sitting on or use your wheelchair tilt and recline to slide you back into place. I bought a female urinal to use on a trip but now use it when I am away from home too long to wait, as well as at home too. It is a real time saver!



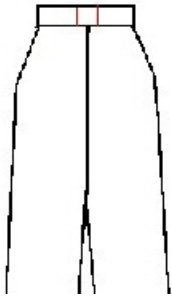
*Two ways to convert your slacks for easy toileting.*

The first method is incredibly simple and, depending on the fabric, may require nothing but a pair of scissors. You need elastic waist slacks that can be pulled up over your hips while you are in bed. This is easy if you are thin and can be done by turning side to side if you are large. Just cut the back seam open (or just cut it out) from an inch or two below the waist band to an inch to an inch and a half before the crotch seam. With sweat pant fabric, that is all you need to do! If simply opening the back doesn't keep the edges out of the line of fire, cut a few inches straight out to the side seams on each side of the opening. Make the cuts at about hip level. Don't cut more than halfway to the side seam or you may have a gap showing bare skin. If the fabric is something that will unravel, use a sewing machine to zigzag stitch next to the cut edges to prevent unraveling. If you make the hip level side cuts, a couple rows of stitching across the end of the cut will help keep it from ripping out farther.

The second method is for slacks without elastic waist bands can be converted as well. Or, if like me, your waist line has expanded since you have been in a wheelchair (That's my excuse and I'm sticking to it!), elastic waist pants large enough to pull easily over your hips may be too baggy

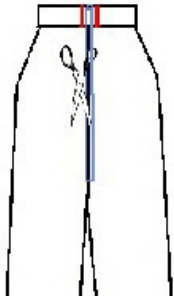
in the legs. It is more complicated to adapt either of these types of slacks, but slacks that fit well are well worth the extra sewing for comfort and looks!

One tip when shopping for slacks to adapt this second way: Waist size is not as important as hip and thigh. The waist will be adjusted when you adapt them. That works perfectly for me since now I don't have to buy pants with clown sized legs just to get them around my belly!



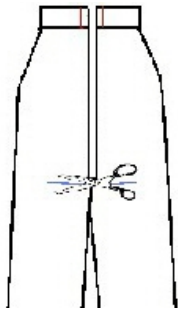
*Back View*

If the slacks you are adapting have an elastic waist band, the first step is to stitch the elastic inside to keep it from retracting when you cut the waist band. Just stitch across the waist band about 1/2 " on each side of the center back seam as indicated by the red stitching lines.



*Back View*

Cut down each side of the center back seam and across the bottom to remove it. Cut through the waist band and down to 1" above the crotch seam.



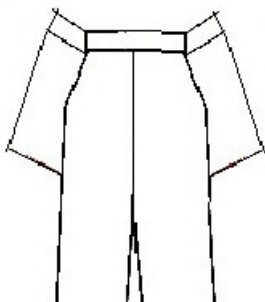
*Back View*

Cut across the pant legs on each side of the bottom edge of the cut out seam. These cuts should be only half-way across the pant leg.



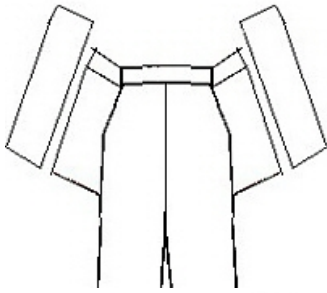
*Back View*

Open the slacks out and zigzag stitch along the edges of the cuts. (A good seamstress or tailor will want to add facings or at least fold and stitch the raw edges. Doing so will put a thick seam directly under the bones you sit on. It will be uncomfortable at best and may lead to pressure sores!) Add two or three rows of stitching outside the point of the cuts to prevent tearing.



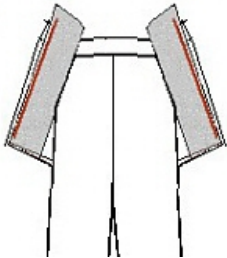
*Front View*

Turn the slacks over...



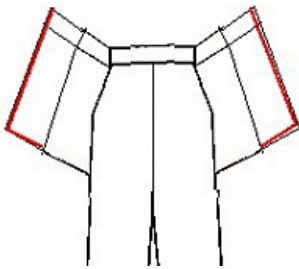
*Front View*

...and cut two pieces of fabric at least four inches wide and as long as the back opening plus enough extra length to fold over to create an extension to the waist band. A thin person may need the strip narrower. It is nice to find fabric that is a close match to that of the pants, but not critical. These flaps won't be visible when you are in your chair anyway.



*Front View*

With right sides together, stitch the flaps to the pants, the bottom edge even with the cuts across the pant legs.



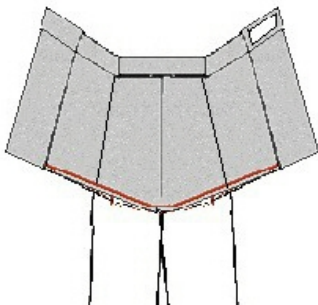
*Front View*

Fold the extra length down to the inside form the waist band extension and stitch. Zigzag stitch or hem the raw edges. (You won't be sitting on these edges so hemming is ok.)



*Front View*

Stitch a strip of Velcro to the outside of the waist band on one flap.



*Front View*

Stitch a strip of Velcro to the inside of the waist band on the other

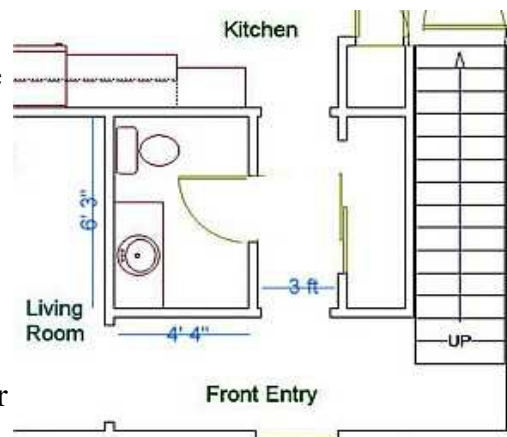


## *An Accessible Half Bath*

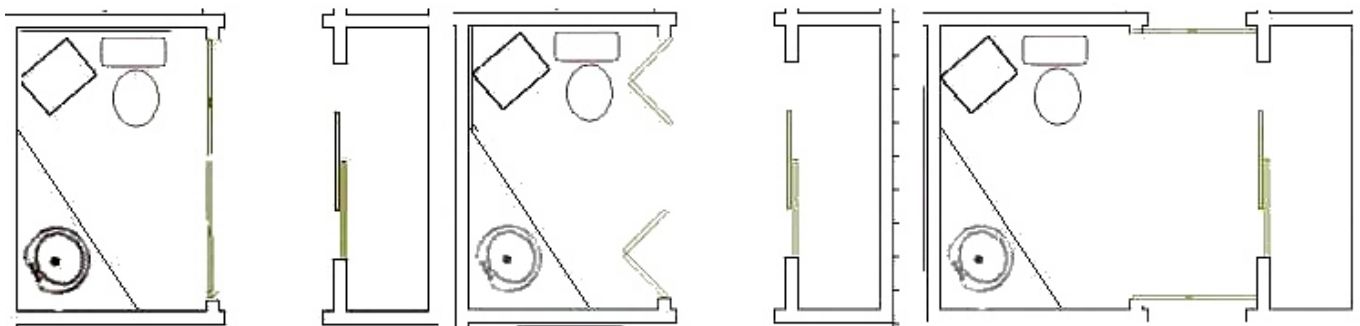
When the process of getting me from wheelchair to chair lift in our small entry hall was getting difficult, it was time to move our bedroom down to the family room. However, standing transfers were getting risky, getting me from my scooter in the hallway, through the half bath door, turned around and lowered onto the toilet was just not safe either. Since it was the only bath on the main floor, we had to figure something out. The cost of adding on a bathroom was beyond our means. I wasn't ready to accept using a commode all the time nor give up on shampooing at the sink. We had a bed shampoo tray that worked well but it was time consuming to use.

I was, however, perfectly willing to give up on showering. Even though I could shower in our upstairs bathroom by using a transfer bench, I had long since switched to daily wash-ups and increasingly infrequent showers. The whole process wasn't worth the effort and extra time. Too many transfers, too shivering cold for me and too sweaty hot for my husband. A daily wash, sometimes just pits and bottom, other times a full scrub down, while I sat on the toilet and a shampoo at the sink was fast, easy on both of us, and more than sufficient to keep me clean. Although a roll in shower would have been nice, it wasn't critical.

So, with a roll in shower off our list of must-haves, the possibility of using the half bath was there - if we could figure out how to get me into it. With the goal of making the toilet and sink usable, we came up with what has turned out to be a very good solution that didn't require major remodeling or changes to the layout of surrounding rooms. This is the "Before" floor plan:



Simply widening the doorway wouldn't do the job. So we W-I-D-E-N-E-D it to 72" to accommodate double doors. Regular doors would be in the way of driving the wheelchair in. The key to success was using 2 two bifold doors. Instead of hanging them from a track, we put them on hinges like a standard door. They fold and swing! When open, they close off both ends of the hallway, turning hall space into bathroom space, nearly doubling the size of the bathroom

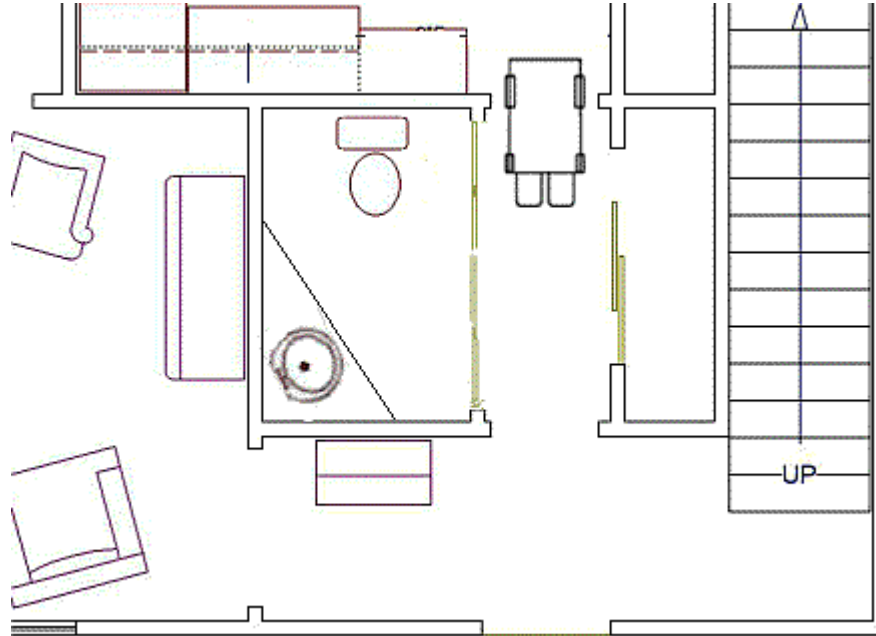


Since we have a basement under the bathroom it was quite easy to move the toilet about a foot away from its original position. This allowed my wheelchair to be along side the toilet for transfers. Removing the vanity and putting the sink in a corner mounted counter top made it

possible for me to roll under the sink for shampooing, etc. Even though I am now on a vent, I can still lean forward over the sink.

We added a tall cabinet in the corner and a cabinet over the toilet for storage. Because the hall closet becomes part of the bathroom, we use half of it for my clothes. A shelf unit holds sweaters, shoes, socks, underwear, etc.

Here is how my power chair (about 46" x 25" with foot rests up a bit) fits in the space.



# **Feeding Tubes**

## ***Why get a Feeding Tube?***

ALS can start with swallowing problems or swallowing problems can occur later. Choking on food or liquids begins intermittently and progresses to inevitable. The immediately life threatening problem is choking badly enough to block the airway. A slower but potentially fatal problem happens as the muscles needed to swallow safely weaken, and food and liquids are aspirated into the lungs causing pneumonia.

Without a feeding tube, the process of living with ALS changes to the process of dying from it. For some people this is fine. They have decided that they do not want, cannot accept, cannot afford, cannot deal with life with ALS. That is absolutely your decision to make! But I can tell you with absolute certainty that a decision made before the reality of swallowing problems are experienced is far less a decision than a wish. As with all the decisions to be made in the course of ALS, you simply cannot know how you will feel until you are at the point where a decision has to be made. That doesn't mean the issue of a feeding tube can or should be ignored until the decision has to be made. A responsible ALS patient will educate himself or herself about feeding tubes for ALS well in advance of the need so that an intelligent choice can be made. It is important to note that a very large majority of people with ALS report that getting a feeding tube was a good decision and that they would recommend it.

At the same time quality of life deteriorates. Dehydration causes low blood pressure, dry mouth, and adds to problems with lung congestion and constipation. Meals are long and drawn out from difficulty chewing, exhaustion, and frightening choking episodes. Both patient and caregiver dread meal times and fear that each choking episode may be the last. Food choices become limited, and eventually even blenderized food is dangerous. Weight loss is significant.

There is no need to experience all the fears, frustrations, and misery of swallowing problems. In the past, most ALS patients lumped having a feeding tube in the same category as going on a ventilator. Today most patients recognize that a feeding tube is a simple, inexpensive, minimal care way to add months, if not years, to their life and quality of life. It reduces stress for everyone, can be used only for supplemental feedings, pills, and extra water and only gradually stepped up to full use when swallowing is not safe at all. And it can be removed at any time you don't want to continue with it.

## ***When to Get a Feeding Tube***

When choking first begins it is time to have a swallow study done. You will be given various textures of food to chew and swallow. The food is mixed with tasteless white goop so that as you swallow, all the stages of swallowing can be seen on X-ray. A therapist can then see how bad your swallow is and recommend ways to make eating safer; chin down to swallow, thickened liquids, double swallow each bite, blenderizing meats.

Even if your swallow study shows minimal swallowing problems, it is time to begin considering if you will use a feeding tube. ALS will progress and swallowing will become hazardous. There really is no point in waiting. Caring for an unused feeding tube is a simple matter of a quick wash of the stoma with soap and water and putting a few ounces of water down it daily. Many people have trouble swallowing pills when swallowing problems first start. Crushing them

and putting them through the tube makes life easier. When you can no longer feed yourself, a can of formula is a time saver. Breakfast from a can really helps shorten the time and effort of getting you up in the morning. And you can continue to eat normally until that becomes unsafe. Many people begin by using the feeding tube for quick meals and supplemental feedings. Later the tube is the main source of food and eating is for pleasure.

When to get a feeding tube is generally thought of as being determined by the onset of swallowing problems. While that is true, the degree of breathing problems is also very important in determining when to have a feeding tube put in. The usual recommendation is to have the feeding tube put in when your breathing capacity falls to around 50%. Even though you may not need the feeding tube for food and water yet, putting the tube in early greatly reduces problems. A breathing crisis during the procedure is unlikely and you will be all around healthier if it is done before you have trouble eating and drinking.

The feeding tube can be put in later but problems are more likely. Placing a feeding tube when breathing problems are advanced is complicated by difficulty keeping the patient well oxygenated while under anesthesia and, if intubation and a vent are needed, it may be difficult to get off the vent afterward. Another problem with delaying feeding tube placement happens as the diaphragm, the muscle that stretches across the bottom of the rib cage, weakens, allowing the stomach to move upward under the edge of the rib cage. That makes positioning the feeding tube through the abdomen into the stomach more difficult. Poor nutrition is debilitating making the procedure harder on the patient and increasing the risk of respiratory problems as well as infection.

Waiting until there is significant weight loss causes even more problems. Not being able to eat enough is slow starvation and reversing starvation is no where near as simple as getting food into them again! When starvation, even slow starvation occurs, body chemistry and metabolism changes. Cardiac problems are common. Refeeding Syndrome will happen when attempts are made to begin feeding. The person will be unable to digest the feedings, be nauseated, feel full too quickly, and may have abdominal pain. ***They are unable to tolerate feedings regardless of the brand of formula or the blenderized food given.*** The changes will need to be gradually corrected with supplements of a surprisingly wide variety of vitamins, minerals, and specific metabolites. This will require the care of a physician familiar with Refeeding Syndrome, generally a specialist in anorexia. Repeated lab work to monitor levels, prevent overdosing, and generally guide the process of refeeding is necessary, possibly daily at first.

## ***The Procedure***

Although we often refer to a feeding tube as a PEG, technically the tube is a “feeding tube” and “PEG” is just one of three possible methods used to put the tube in place:

If you look up “feeding tube” you will see pictures of people with tubes in their nose. Ignore these. They are for temporary use and are not for people with ALS.

- PEG: Percutaneous (*through the skin*) Endoscopic (*inserted using a scope down the throat and into the stomach*) Gastrostomy (*an opening through the stomach wall, abdominal muscle, fat layer, and skin*).
- PRG: Percutaneous (*through the skin*) Radiological (*using x-ray radiology to visualize the stomach*) Gastrostomy (*an opening through the stomach wall, abdominal muscle, fat layer, and skin*). Also known as RIG.
- Surgical Gastrostomy, done in surgery.

Another feeding tube option is a J tube which can be placed by either PEG, PRG or surgical method. For this, the feeding tube is placed in the jejunal section of intestine just beyond the stomach. Since the stomach is mainly a blender that liquefies food before it moves into the jejunum, the digestive process isn't disrupted. This type of tube is used for people who have problems with gastric reflux (food moving backwards from the stomach into the esophagus). That causes heart burn and scarring of the esophagus. The potential for that liquid food to move upward far enough to get into the lungs is a concern especially for any ALS patient who can't sit up after tube feedings.

Either way, a tube is placed through a tract through the abdominal wall so that liquefied food can be poured directly into the digestive tract rather than being swallowed. Regardless of the method used, the tube itself will be chosen from several similar brands. Rather than calling it a PEG, it should be referred to as a G tube or J tube, technically accurate regardless of the procedure used. Feel free to call it a feeding tube, G tube, J tube, Harold, Eloise, or another name of your choosing.

Breathing is a big concern during this otherwise minor procedure. For respiratory support he may use BiPAP or, if your respiratory status is poor, intubation (a breathing tube through the mouth into the trachea) may be needed. Whether or not you will spend a night in the hospital depends primarily on whether your breathing becomes a problem and your doctor's preferences.

The decision regarding which method to use for insertion of the feeding tube will be made based on your breathing status and possibly the availability of a Radiologist who performs PRG's. Although PEG is more common, PRG is now being used more frequently for ALS patients who have a moderate or severe level of respiratory involvement. (Moderate = lung capacity of 30 to 50%, Severe = less than 30%) It is especially helpful in cases where the weak diaphragm (the muscle that separates the abdominal organs from the chest) has allowed the stomach to move further upward under the rib cage.

The PEG procedure is generally done in the Endoscopy Department of the hospital. An Endoscopic tube is placed through the mouth and into the stomach. The camera of the endoscopic tube allows the inside of the stomach to be seen. The PRG procedure is done in the Radiology department with radiography (live x-rays) being watched during the procedure. The x-rays allow the stomach and other organs to be seen.

For either procedure the patient will have an IV for medication and be sedated. BiPAP can be used during the procedure and breathing will be monitored. The sedative may be given with pain medication for deeper relaxation and sleep. The amount of sedation/pain medication for the PEG procedure is generally higher because the endoscopic tube is quite large. For the PRG a much smaller and softer tube is put through the mouth to the stomach.

After sedation the Endoscopic or smaller tube is inserted. From this point, the procedures are very similar. The direction to best enter the stomach is identified and the area on the upper left abdomen is numbed. A guide wire is used to make a path from the small abdominal incision to the stomach. The feeding tube is pulled along the same path by the guide wire. The guide wire is then removed by pulling it up the endoscopic tube or the smaller tube and out the mouth. Done!

Immediately after you wake up after the tube is placed, you will probably want some strong pain medication. By the next morning, the pain should be considerably less but you will still want some kind of medication. Tylenol will soon be enough. Because placing the tube requires going through abdominal muscle, pain when moving will last several days. Using a pillow to support your abdomen when lying on your side in bed will reduce pain.

## *The First Tube*

If you are able to meet with the doctor performing the procedure before the feeding tube is put in, ask to see the type of tube you will have. If not, ask the nurse afterwards. Knowing what the whole thing looks like, inside and outside parts, is reassuring in its simplicity and the knowledge that you don't have to be an engineer to understand how it works and how to troubleshoot problems.

The first feeding tube may be a long, opaque, rubber-like tube, a clear soft plastic tube, or it may be a clear soft plastic that extends only about six inches out from the skin.

The first tube is often held in the stomach by a flat or mushroom shaped disk inside the stomach rather than a balloon as may be used for replacement tubes. Further up the tube, on the skin, is another bumper or pair of plastic wings. This bumper encircles the tube but isn't attached to it so that it can be slid up or down the tube. It has a firm grip on the tube though, and won't slide by itself. The purpose of this bumper is to stabilize the tube and to keep the tube from being sucked into the stomach by normal digestive action. The inner bumper or balloon would then block the flow of food into the intestines.



The first tube may be a single tube or may contain another separate tube inside it. At the top of the tube are one or two ports. Note: These are the most common configurations for the first tube, but there are other types. Again, knowing which type you have and being given the package it came in, complete with an illustration of the tube, is very helpful.



A single port tube is used when the tube feeding will be given slowly from a bag. It can be run through a pump that will deliver a precise amount of feeding per hour. This is handy for patients who don't tolerate getting a large amount of feeding at once. A low tech way of doing this is to use a clamp on the tube to adjust the flow to a rate the patient can tolerate.



If the tube has two ports, the larger one has a tab top that is opened for feeding. A large 60 cc syringe is used for this port. It has a tapered tip the fits into the port. The fit isn't one that will grip the syringe so I strongly recommend holding the syringe and port together with one hand while pouring the feeding. Using the plunger in the syringe is totally unnecessary. It increases the risk of blowing the feeding all over, or giving the PALS a stomach cramp, and isn't significantly faster. Just use the syringe as a funnel and let the feeding run in by gravity. If it doesn't flow well, either the liquid is too thick and needs water added, or the PALS needs to recline more for the feeding.

The other, smaller port is for medicines. Medications can easily clog the small medication tube and are nearly always given through the larger feeding port. The only use for the medication port is to give meds if the patient has a continuous drip feeding. It saves the messy process of opening the running feeding port to add meds. The medicine port will only attach to a syringe that screws in, called a Luer lock syringe.

In my experience, much of the pain I experienced was due to the tightness of the outside bumper on the tube. Surgeons push these down against the stoma (opening through the skin) very

tightly and I can only guess that the reason is to pull the inside bumper or balloon snugly against the stomach wall inside. This pressure would help prevent bleeding from the hole in the stomach wall. Unfortunately, what is snug on the inside puts painful pinching pressure on the skin. The outside flange digs into the skin and even looks painful. This is easily and safely fixed the day after surgery. The flange is a soft plastic ring, triangle, or pair of wings that is not actually attached to the tube. To back it away from the skin, simply lift the opposite edges of the flange and pinch them up against the tube. This will open the hole in the flange and allow it to be slid along the tube. It may stick at first and you need to be careful not to tug on the tube, but with gentle manipulation, it will move and the pain relief will be nearly instant.



The combination of anesthesia and pain meds will almost inevitably cause constipation if a stool softener (Dulcolax, Colace, or Docusate) isn't taken daily, ideally starting a day or two before the feeding tube is put in. It is critical that you not begin eating or being fed through the tube until your intestines "wake up" from any anesthesia you are given during the procedure or surgery. For some reason, intestines are slower to shake off the effects of anesthesia than the rest of the body. *This applies to any procedure or surgery!* There is some evidence that gastric motility, the passage of food through the stomach and intestines, is slower in ALS patients, but whether this is a direct effect of ALS on digestive muscles is far less likely than the effects of not being able to get up and walk! If you ever had surgery before ALS, you know that you were ordered up and walking far sooner than you wanted to. The assumption is that it was to help you get your strength back. Not really. It was to get you breathing more deeply and get your bowels working.

If you are in the hospital during this time, your nurses should be listening to your lower abdomen with a stethoscope to hear the gurgling that signals the return of bowel function. Until then, you must not be fed. Severe problems with the intestines can occur in anyone after surgery and because of immobility, ALS patients are somewhat more susceptible. If you have the tube placed in an outpatient procedure and go home after or are only kept overnight, you probably haven't been given heavy anesthesia and shouldn't have this problem, but waiting to hear bowel sounds before beginning eating is still important. A stethoscope is handy but just putting an ear to your lower abdomen will let a caregiver hear the gurgling. Once you are being fed, if you haven't had a normal bowel movement by three days, it is time for a laxative. If no results in twenty-four hours, repeat the laxative. Let your doctor know if you aren't able to go by five days. Don't mistake passing liquid stool for a bowel movement. When constipation becomes serious, only liquid stool can get past the hard stool blocking the intestines.

## ***Stoma Care***

The skin opening for the feeding tube is called a stoma. The name has nothing to do with the word "stomach" in spite of the spelling. The word stoma refers to any tunnel through the skin and underlying tissue to a place inside the body. Stoma can refer to the opening of a tracheostomy, colostomy, urostomy, and others.

## ***Stoma Cleaning***

If you are told to clean around the tube with peroxide, stop using the peroxide after about a week. It is great for breaking down dried blood and gunk and foaming it away, which is why it is commonly used after any surgery. However, continued use also breaks down the "scaffolding" of



blood clotting needed for wound healing. The constant disruption of the attempts to heal cause the body to try harder by producing even more healing tissue which is called granulation tissue. This becomes chronic around the feeding tube and repeated bouts of painful, burning, bleeding granulation tissue occur. So stop using the peroxide. It is not effective for preventing infection anyway. Washing once or twice daily with soap and water will do just fine. There is no longer a need to make washing it a sterile procedure (sterile gloves, sterile water, and q-tips) by this time. Even so, care needs to be taken: Use a different wash cloth than the one used for the rest of your bath. A thin, cheap or worn out wash cloth gets under the bumper much better than a plush expensive one. Antibiotic soap is not necessary (and environmentally bad as it ends up in our water supply). The amount of antibiotic and the time it is on the skin is ineffective anyway. The soap itself has just as much antibacterial action. A split gauze 4x4 or 2x2 under the flange plate will catch any drainage and keep the plastic from irritating the skin. At first Bacitracin ointment should be applied with a q-tip, but can be skipped after a few weeks.

### *Stoma Problems*

Feeding tube stomas (openings) never heal completely so you do need to watch for signs of infection. Infection doesn't usually stay just red for long. It gets increasingly sore, pus begins to drain around the tube or the skin deteriorates to a raw oozing sore. Often there is a bad odor or a fever which may be low. In case of infection a prescription ointment, not a simple over the counter like Bacitracin will be needed.

You can easily tell the difference between infection and granulation tissue. Infection will cause the entire stoma to be red and sore. Granulation appears as raw, red bulges on the stoma itself that bleed easily. Granulation tissue will shrink to nothing in a day (possibly two) by applying a dab of non-prescription hydrocortisone cream. Don't use it daily, just when the stoma has granulation tissue. Doctors are fond of using silver nitrate sticks to burn the granulation away. Ouch. You can get these for home use. There may be some reason why doctors prefer silver nitrate -- they can't all be sadists -- but I have never heard the reason.

The occasional brownish drainage is pretty normal, as is some dried blood or even fresh blood. Worry only if it is saturating the gauze and won't stop.

Trying a number of things may be necessary for skin problems. You can speed up the process if the red area is big enough to try one thing on half and another on the other. If the redness is irritation from the gauze Vaseline will help. If it is itchy too, try a cream made for athletes foot. Cortisone is great for granulation tissue but that is usually right on the stoma, not the skin. Cortisone is soothing for other causes even if not a cure. Mylanta or a paste made from crushed Roloids and water will neutralize any leaking stomach acid. A diaper rash ointment is a good moisture barrier. And sometimes nothing but a bare belly exposed to the air works, especially with sunshine.

### *Tubing Care*

When you get the first tube and with each change, ask to keep the package the tube came in. This will help with replacement of the same type and size of tube if it should come out. Write the date on the package.

When you first get a feeding tube, have one replaced, or slide the bumper back, make a note of the position of the bumper on the numbers along the tube for comparison if problems occur. Write this on the package too.



When your feeding tube is first placed or replaced or with any abdominal pain, checking to make certain that the tip is in the stomach is critical. Running tube feeding into the abdomen rather than the stomach causes **severe** complications. So, with the first feedings, you will be taught to check tube placement before feeding. Any pain during these checks beyond the discomfort of having the tube moved around, indicates trouble requiring a call to the doctor. The pain will be bad and there will be little doubt there is a problem.

- ✓ Begin by checking the position of the outer bumper. The number closest to the stoma opening should match that which you so carefully recorded the last time the feeding tube was put in or the bumper adjusted.
- ✓ Next, use the big syringe and its plunger to draw back and see if you get stomach contents. If so, you are done checking and can go ahead with the feeding. If the stomach is empty you won't get enough, if any, stomach contents to be sure, so additional checks are needed.
- ✓ Put your ear on the person's belly or use a stethoscope to listen for a whoosh and gurgle as you use the syringe to push some air into the stomach. If that checks out it means the tip is in or near the stomach. "Near" isn't good enough so do the next step.
- ✓ Use the plunger to push some water in and then try to pull it back out. If you use cold water the person may feel a cold sensation in the back of the throat because the sensation is transmitted there, not because the water is going up there. If this step checks out without sudden pain, you can go ahead with the feeding.
- ✓ The only better check is an X-ray. This will probably be done if when you have are having the tube replaced but certainly isn't necessary before every feeding.
- ✓ If the person has a trach as well as a feeding tube, be certain that every caregiver is alert to the risk of mistaking the balloon port on the trach tube for the feeding or medication port on the feeding tube. The ports can look very similar, right down to the color. Feedings given during the night may be hooked up without turning on full lighting and be attached to the wrong port. Really unlikely, but stranger things have happened.

How long do you have to keep doing this song and dance every time you use the feeding tube? No one will give you a straight answer to that! My best answer is until you have done it enough times without any sign of trouble that you are comfortable skipping it. If you have home nursing care, be prepared for this ritual to go on forever. Most nurses are taught feeding tube care based on the nasogastric (through the nose and down to the stomach) type of feeding tubes that are far more common in hospitals than feeding tubes. Nasogastric tubes can easily be tugged up out of the stomach by a patient or even cough/gagged upward. If that happens there is a real risk that a feeding will end up in the lungs. Bad thing. So, nurses with hospital experience have the need for ongoing tube placement checks burned into their brains and this may follow them into home care even though feeding tubes aren't as problem prone as nasogastric tubes. Once in, a feeding tube isn't going to go anywhere unless it is pulled hard. If that should happen, of course, you will want to check placement before using it again.

The Number One rule for caring for a feeding tube is to flush it with at least two ounces (60 cc's) of water every time you use it and once a day if you are not using it. Unless you do that religiously, even fanatically, the tube is going to get plugged up. To flush the tube, use the 60 cc syringe as a funnel. Don't use the plunger to push the water through, allow it to flow in by gravity. You will quickly become accustomed to the speed at which the water will flow in and can tell if the tube is gradually clogging up.

A plugged tube may have to be replaced, but usually can be unplugged if attended to promptly. If you are having problems with a newly placed tube, it probably isn't simply clogged if you have been flushing it well. This situation requires a call to the doctor. However, in a tube that has been working well, check the markings on the tube to make sure it is still in the stomach. If that checks out, try these steps:

- ✓ Because the rubber/plastic is soft you can attempt to dislodge a clog in the external section of the tube by squeezing the tube as you move your fingers down it towards the stomach. A little lotion on your fingers or the tube will help.
- ✓ Using the syringe, push about 20 cc's of air forcefully through the tube. If it won't go in, the clog is a big chunk and will need to be dissolved. Sometimes it will soften just by filling the tube with water and letting it sit for a few hours and then using the syringe to push water through.
- ✓ Another possible fix is to put Coca Cola in the tube overnight. Urban legends abound about the harshness of Coke, but this does seem to work!
- ✓ Use a feeding tube brush or pipe cleaner. Pipe cleaners of the type used for crafts are too soft to work. Real pipe cleaners from a tobacco or pipe shop are stiff and work well if you can find the right size. If you use a pipe cleaner, don't push it in past the point where the tube goes through the skin. Brushes can be found online by doing a search for tube feeding brushes. The info may say it will work with a 20 French tube or larger. Since it would be very rare for an adult to have a tube smaller than a 20, this brush will work.

Positioning a feeding tube in any particular direction, if at all, is a matter of choice. The lump under your shirt is certainly more apparent to you than to any one else, but most of us feel the need to minimize it. There are tube holders you can buy but you can make one from a strip of soft fabric and Velcro. A length of Ace stretch bandage like you use for a sprained ankle works well. Taping the tube in place is not comfortable, and replacing the tape daily causes tape burns. Whichever way you do it, put a Kleenex between the tube and your skin. Maybe it is just me, but the ports on the end of the tube sometimes give me a blister.

If the look of the tube really bothers you, a button feeding tube can be put in at a tube change. This tube is capped at the stoma, eliminating the dangling length of tubing. The price you pay for this little vanity is that using the tube requires attaching a short length of tubing which adds steps to the feeding process, but may be worth the trouble in trade for restoring your self image.



You may see a build up of black stuff in the tube. This is yeast, which finds the warm, wet interior of the tube to be yeast heaven. As nasty as it sounds and looks, yeast growth is harmless. Any of it that gets washed into your stomach is quickly killed by stomach enzymes and acids. Regularly cleaning your tube with pipe cleaners or feeding tube brushes keeps it from looking really bad, but the best they can do is remove some of it. Even if you were able to get the tube looking spotless again, studies have shown that yeast embeds itself invisibly in the tubing material and quickly re-grows.

### ***First Tube Replacement***

A tube with a bumper in the stomach can last for many years before replacement is needed. Rubber tubes will deteriorate very slowly but the plastic ones don't seem to deteriorate. Rubber tubes will become lumpy as they deteriorate. The usual reasons for replacing the original tube are clogging problems or the annoyance of having the little tab that plugs the tube break off.

The tube is generally replaced by a typical feeding tube. These do not have the disk to hold it in place but instead have a small balloon. These tubes generally have three ports; a large port for the feeding, a small port for medication, and a medium sized port for air or water to fill the balloon. The balloon port doesn't have a cap but is filled using a screw in tipped syringe.



With the first feeding tube change you can request any type or brand of tube you want. Your original tube was most likely held in your stomach with a bumper (also called a disc, flange, bolster, mushroom tip). They won't fall out and would require a very hard tug to pull them out. Tubes with bumpers require less frequent changes, but because of the potential for damaging the track through the abdomen by pulling the disk out through it and the risk of bleeding, especially if the patient is on Coumadin, most physicians prefer to remove the tube endoscopically. This requires light anesthesia (twilight sleep) to put the endoscopic tube down the throat into the stomach. BiPAP may be used. The disk is clipped off and pulled back up through the throat and the feeding tube is slipped easily out of the abdominal opening.

The other type of tube has an inflatable balloon in the stomach to keep it from sliding out. Because the balloon deteriorates from stomach acids, it begins to deflate, usually at about six to nine months. When the balloon begins to deflate, the tube can be accidentally pulled out with a tug. With further deterioration of the balloon, the tube will simply slide out. If the tube comes out, it needs to be replaced within twelve hours at the very most before the track collapses and closes. That will require a new PEG or RIG procedure to reinsert it. The original stoma may be used but a new track through the abdomen and into the stomach is generally needed. Your doctor will prefer to change it every 6 months rather than have to do an unscheduled, emergency change if it falls out.

A popular tube is a low profile button tube. These are commonly Mic-Key tubes although there are other brands of low profile tubes. They are available in bumper or balloon type. This type has no tube dangling out so the risk of pulling it out is reduced. An adapter between the button and tubing or syringe for feeding is required. The adapter will make it harder for a patient with weakening hands to do his own feedings, but for most caregivers, it is just one small extra step in feeding. One big advantage for some patients is in self-image. There is no tube or clamp to show as a lump under clothing.

Changing and replacing balloon tubes is simple and painless. The balloon is deflated and the tube is pulled out with only a slight tug. The new tube is lubricated with KY gel and inserted and the balloon inflated. This can be done by a doctor, or, after the tube has been in a year or so and the track through the abdomen is well established, it can be done by a nurse or caregiver. These balloon tubes do require more frequent changes but are much easier to change, especially for patients with breathing problems but not on a full ventilator.

### *If the Tube Comes Out.*

One sign that the balloon is shrinking and getting stiff and may fall out is an increase in drainage around the tube as the balloon shrinks and doesn't block the path around the feeding tube as well. Tube feeding formulas will leak a tannish brown gunk. There will be no pain, tenderness, redness or bleeding. The tube will also get looser and slide out further as the balloon deflates. You can baby it along for a while by pushing the tube back in and adding air to the balloon, but that quickly becomes a daily need. Take extra care not to let the tube get tugged on. Taping it down can help assure that the tube stays in until your tube change appointment but if you delay too long the balloon won't hold air at all. Then the feeding tube can fall out and you will have to have it

replaced within about 6 hours -- twelve is considered the outside limit -- before the tract to the stomach closes off too much for easy replacement. Begin by calling the doctor who put the tube in or your Home Health Nurse if you have one. If they are not available or the nurse is not allowed to reinsert it, just go to the emergency room and they will replace it. After watching a couple of simple replacements your caregiver can do it!

### ***What's for lunch?***

Adjusting to tube feedings is an experience that is extremely variable. For some, the feeding tube is put in before swallowing problems make eating dangerous. For them the tube is an easy way to get medications down and get plenty of water. Supplemental feedings through the tube help maintain weight and nutrition. A meal of a tube feeding can be a real time saver for a caregiver. I have a can in the morning since that is the busiest time for my husband caregiver. Not having to fix and feed me breakfast on top of getting me washed, dressed, and beautified to face the day is great.

The choice of what to put down the tube depends on several factors; caregiver's time, digestive problems, medical conditions, insurance coverage. Very few tube feeding formulas have any dairy product in them because so many people are lactose intolerant. They may not know it until they begin consuming more dairy products in order to maintain weight.

Boost and other products sold in grocery stores are formulated to be supplements to a regular diet or for short term use as a substitute for eating. "Real" tube feeding formulas are able to provide all the nutrients needed to replace a normal diet and need not be for short term use only. These are unflavored and not intended to be swallowed, only used through a feeding tube.

Top brands such as Jevity have different types to meet the calorie requirements of individual patients. The higher the amounts of calories, protein, etc. the thicker the formula and the need for more free water. Free water is plain water with nothing added. Anything added requires a portion of the water to be used to flush the ingredients from your body, so coffee, tea, soda should not be considered as good fluid intake.

Nearly all contain fiber for bowel regularity. There are also specialized formulas for diabetics, people with liver, kidney, or lung disease. Your doctor or a dietitian or nutritionist can recommend a brand but be prepared to change brands if one doesn't work well for you.

The big question is always "Does Medicare pay for tube feeding formulas?" Yes, the formulas and supplies are covered by Medicare Part B and most insurance.

- Formula won't be covered unless it is the primary source of nutrition and you can't eat well enough to maintain your weight without it. Medicare covers tube feeding formulas if they provide all or the majority of your nutrition. Being able to swallow small amounts of specific foods doesn't disqualify you, but if you still eat most of your meals and just use the tube feeding as a supplement or convenience, Medicare won't cover it.
- A form/documentation by your doctor is required for Medicare reimbursement and a diagnosis of ALS covers all the requirements.
- Standard tube feeding formulas are covered as well as special formulas needed by diabetics, kidney or liver patients, etc. (Do an internet search for Medicare Part B Enteral Nutrition Reimbursement Manual.pdf. It lists many but probably not all covered formulas.)
- Natural formulas may be covered if your doctor documents an allergy or intolerance of standard formulas. A preference for a natural formula isn't covered.
- The cost of foods and supplements used to make your own blenderized formula is not covered.

If you get formula through a Medicare provider, compare the amount of your copay against the price you would pay buying it yourself online -- no prescription needed. Often prices are so jacked up by Medicare approved providers that you end up paying more copay even after Medicare limits the amount Medicare will pay.

Many people opt to continue using regular food by blenderizing it. Some feel this makes them feel more normal while others believe it provides better nutrition. Whatever the reason, a proper balance of nutrients, not the latest health food fad, is necessary and this requires a cook who is knowledgeable or willing to learn. Eating what the family eats may be fine, but it does depend on how much time the caregiver has to devote to meal preparation. Caregivers are often overwhelmed and exhausted as care needs increase. It is important that both the person with ALS and the caregiver be open to using prepared formulas partially or completely.

Digestive problems when beginning tube feedings aren't the norm but do occur. Common problems are diarrhea, bloating, cramping, or nausea with feedings. There are a number of formulas available and a dietitian will evaluate you and suggest a formula and amounts. If there are problems, the dietitian can make changes until a solution is found.

Often the solution to tube feeding digestive problems is an adjustment in how the feeding is given. Cold feedings cause cramps. (Putting the next few cans on top of the refrigerator brings them to a comfortable temperature for me.) The first feedings need to be small and given slowly as tolerance increases. Even later, too much feeding at once causes problems. Some people can tolerate a couple of cans at once, but many, especially those not yet on a vent or using BiPAP after eating, have problems with big volumes. Using the plunger to push feedings in is not necessary or comfortable. Use the big syringe as a funnel and let it flow in by gravity.

Some people do better with feedings given at a slow drip. The feeding can be given from a bottle or a bag suspended from a pole, hook, chandelier, or on a special pump to precisely measure the drip rate. That is seldom necessary because the tubing from the bag has a clamp that can regulate the drip rate. Running this type of feeding overnight with an occasional "snack" during the day can free you from the tube and make moving around easier.

Feedings should be given sitting up just as you would normally eat a meal, and you should remain sitting up for at least a half hour after eating. Heart burn (esophageal reflux) is no more common with tube feedings than regular food, but sitting up will help prevent it in people prone to heart burn. If you have home nursing care, you will once again find your nurses applying nasogastric tube feeding rules. A nasogastric tube passes through the sphincter (muscular valve) between the esophagus and stomach. The tube can keep the sphincter from closing completely and allow the feeding and stomach acids to leak back up the esophagus, especially when lying down. So a nurse won't let you lie flat, even you never get heart burn and could take your feedings standing on your head!

Too often problems that are attributed to tube feedings are no such thing. Bloating, gas, abdominal distension, nausea, and vomiting are all signs of constipation. Unrecognized and untreated this can lead to a bowel obstruction, a serious problem often requiring hospitalization.

This problem is especially common in the week following having the tube put in. Any anesthesia puts the bowels to sleep and they can be slow to wake up and resume the muscle contractions that move food along the digestive tract and turn it to fecal matter and cause a bowel movement. If you are one of the lucky few who escape from the hospital within hours of having the feeding tube put in, it is extremely important not to begin tube feedings until the bowels are back at work. Use a stethoscope or put your ear to the persons belly and listen for gurgling. It won't be a constant sound but should be there. That indicates all is well and slow, small feedings can begin.

Passing gas is another sign that the bowels are working, and actually having a bowel movement is cause for great joy and celebration!

Prescription pain meds are also notorious for causing constipation. That doesn't mean you have to suffer, but you should take extra precautions. As mentioned earlier, a stool softener (some form of docusate sodium, available without a prescription) started before you go in for your feeding is very helpful. It prevents the stool from hardening too much as it moves through the bowels. It is available as a liquid but the pharmacist at the drug store may have a day or two delay in getting it in stock. If you take it by mouth before getting your feeding tube, DO NOT take it straight! Put it in orange juice or other thick liquid or it will burn all the way down. Once your feeding tube is in, you can put the Docusate liquid down the tube.

Until you are well established on tube feedings and having regular bowel movements, keep track of when you last had one.

A little tutorial about the consistency of bowel movements is needed here. Diarrhea and liquid stools are not the same. Diarrhea stool contains brownish water with bits of stool. That is likely to indicate that you cannot tolerate the type of formula being used. It is also common after time on antibiotics. Liquid stool is thicker brown liquid that can be from either of the above cases. More importantly in this situation, it can also indicate constipation or blockage since only liquid stool can make it around the compacted stool in the bowel. Unless the amount of liquid stool is huge and frequent, the cause is constipation with obstruction. It cannot be counted as a bowel movement. Give a strong dose of a laxative. If no results in 24 hours, repeat it, or step up to an enema. If 5 days pass with no real bowel movement or the person won't eat, is nauseated, call the doctor. If vomiting starts, call immediately or go to the Emergency Room, as you have reached Code Brown Concrete Alert and may require medical dynamite.

### *A Little Noted Feeding Tube Plus*

You know those times when the whole family has stomach flu and you are dreading catching it and the nausea, cramping, and weakness that proceeds every bout of vomiting? When you feel that building up, you can skip the worst of it by using the big syringe and plunger to empty your stomach. No barfing!

### *Feeding Tubes and Pills*

Putting medications through the feeding tube is generally not a problem, but there are rules to be followed.

Most pills can be crushed, put in water, and poured down the tube. They have to be well crushed so investing in some sort of pill crusher is helpful. A simple mortar and pestle (bowl with a rounded club) works well. It is hard to find as a medical product -- look for it as a cooking tool. There are many other types of crushers available but if your caregiver has arthritis, you may have to try a couple of before you find a usable one.

Many medications are available in liquid form -- for a price! Simple Tylenol tablets all seem to be coated these days, but still can be crushed with a bit of extra grinding. If you want liquid Tylenol, children's liquid is readily available. It is more concentrated than adult liquid, so make certain you aren't using too much. Adult vitamins are about three times more expensive in liquid form even if available but are increasingly common in chewable form that can be easily crushed.

The problem medications are those that are designed to release slowly into the digestive tract for longer, more even effect. If crushed the entire dose will hit all at once and that is not at all good.

These medicines usually have sustained-release [SR], extended-release [ER, XR, XL], controlled-release [CR], after their name. Often they are capsules with beads of medication inside. If a tablet has a line scored across it for cutting, it can be crushed as can meds labeled as immediate-release (IR) Try to remember to let your doctor know that you need crushable or liquid medication and double check with the pharmacist. See the list of "Do Not Crush" medications at <http://www.ismp.org/tools/>



# *Respiratory Options*

## *Ineffective Coughing*

I remember all too well the hours spent trying to clear my airway with a cough too weak to do the job. Frightening for everyone and so exhausting! Get a Cough Assist machine as soon as possible. A Cough Assist machine mimics a strong, natural cough. A full breath is pushed in through a mouth piece then the pressure is abruptly reversed to a suction level causing the equivalent of a good cough. Quick and easy. The machine can be set to automatic or to manual where the patient times his cough with the machine's inhale and reverse cycle. I don't recommend the manual setting because if you feel really short of breath it is hard to relax and time your cough. Not to mention that someone as uncoordinated as I am may never get the hang of it!



The Cough Assist can be used with a mouthpiece or a mask. It can also be put on a trach. It is portable at 10x11x17 inches and 26 pounds.

Ideally you have a Pulmonologist, and if you don't it is time to get one. A neurologist and family doctor are not the ones qualified and experienced with the breathing problems in ALS. The pulmonologist will likely be the one who sees you through to the end.

While waiting for the Cough Assist machine to arrive, here are some things you can do. There are cough medicines that contain guaifenesin that works quickly to thin the mucus so you can cough it out. Mucinex is a good one, and Walmart may still have a generic brand that is just guaifenesin without other stuff. If you can't find that, look for a cough medicine that says it is an expectorant, such as one type of Robitussin. Regular cough medicines usually have cough suppressants in them as well and we don't really want that.

Some people swear by papaya juice for thinning mucus. Others use the liquid from a jar of pickles! Since I have a trach for suctioning (the absolute best way to get the crud out, but a little extreme if you don't need a vent as well) I haven't tried these refrigerator cures but wouldn't be at all surprised if they worked.

When you feel congested, take a generous dose of the "cough medicine" you prefer. Give it a few minutes to work, then lie down. This will feel like the absolute wrong thing to do! But lying down will allow the mucus to be propelled upward more easily with gravity less a factor. Lying down will also allow someone to help by applying a manually assisted cough. To do this place both hands on the abdomen just below the ribs. This can be done with the hands one on top of the other as is done in CPR, or with the thumbs together in the center and hands out toward the sides. Have the patient take three deep breaths if possible and on the third cough, apply a quick thrust, not just pressure. Direct the thrust upward under the ribs. This will give that "Oof" of having the wind knocked out of you. That will add force to the cough and move the mucus upward. Repeat a couple of times. Rest between assists and repeat until the airway is cleared.

You will also want a suction machine and what is called a "tonsil tip" plastic wand (brand name Yankauer) to help remove the mucus from the mouth if necessary.





You may not have to use the tonsil tip suction wand at all, but just having it ready is a much-needed reassurance for everyone.

Increase water intake to keep the mucus from being thick and sticky. If swallowing water is difficult, make sure it is hot or cold. In between, the water won't trigger a good swallow. You can also try adding a thickener such as "Thick-It" to any liquids to make them just a tiny bit easier to swallow. Water is actually the hardest thing to swallow. Flavored or fizzy drinks may go down better as will milk (not advisable as it can increase congestion) or orange juice. If swallowing liquids is difficult, it is time for a feeding tube. You may not need it yet for nutrition, but good hydration is critical for the lungs. A cup of water down the tube about four times a day helps all body systems including the ever problematic bowels!

## ***Oxygen Use with ALS***

At some point in ALS progression the question of using oxygen to ease breathing difficulty will come up. Getting enough oxygen is not a the problem in ALS. There is nothing wrong with the lungs. The problem is muscle weakness that prevents inhaling deeply enough to get enough air in. If a machine such as a Bi-level Ventilator or an invasive ventilator is used to push enough air in, the lungs can absorb oxygen from room air without difficulty. Therefore the treatment for breathing problems in ALS is mechanical support, not oxygen. For people with ALS who also have lung problems that directly affect the ability of the lungs to absorb oxygen (such as pneumonia, COPD, emphysema or asthma) in addition to ALS, oxygen may be necessary.

Often the response to the use of oxygen is that it is dangerous for ALS patients. That is both true and false! Respiratory drive runs on CO<sub>2</sub> levels. Oxygen levels contribute very little to the process of stimulating breathing. CO<sub>2</sub> is produced by working cells and sent through the blood to the lungs to be removed during the process of breathing. The CO<sub>2</sub> is exchanged for oxygen and the CO<sub>2</sub> is exhaled. When that exchange is impaired, CO<sub>2</sub> levels rise and the respiratory regulatory center in the brainstem coordinates an increase in breathing rate and depth to blow it off. An ALS patient with weak respiratory muscles can't breathe deeper so the CO<sub>2</sub> levels are harder to bring down.

Over time body chemistry allows the respiratory system compensate and to work with these levels as the "new normal". When the compensatory measures are maxed out by increased weakness of the respiratory muscles or lung congestion or pneumonia, that small amount of respiratory drive from oxygen becomes very helpful. But if oxygen is given and the O<sub>2</sub> Saturation increases to about 90%, that part of the respiratory drive stops because the O<sub>2</sub> level is near normal.

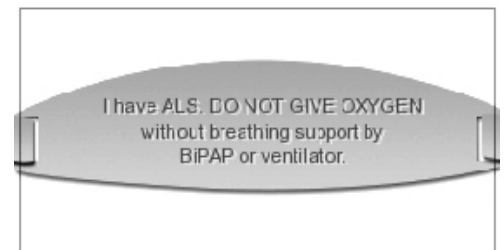
Loss of that small part of respiratory drive is enough to tip the drive from "barely enough" to "not enough". The rate of failure begins to increase and the patient may stop breathing entirely. The

The risk of giving oxygen to an ALS patient who has high CO<sub>2</sub> levels is not recognized by some doctors and respiratory therapists. If you develop breathing problems and call for an ambulance or go to the Emergency Room, protect yourself by wearing a Medic Alert bracelet. They are available online from several companies.

This site offers a range of styles:

[picturesongold.com](http://picturesongold.com)

The suggested wording is shown.



risk of adding O2 is therefore very real -- but only if the patient is retaining CO2 enough to rely on compensatory body chemistry.

So when does this risk begin with ALS? It is a basic rule in medicine that you don't automatically give more than 2 liters per minute of oxygen to anyone. This amount is considered low enough to be safe for anyone, but a check of an ALS patients CO2 levels should be done regardless. O2 Saturation (monitored with a simple fingertip monitor) does not reflect CO2 levels and O2 levels remain normal until CO2 levels are very high. The person's CO2 levels have to be checked to see if he is retaining CO2. This can be done using a device that tests exhaled air (Capnometer), or with a check of blood drawn from a vein, the same as for most blood work. The results of these methods correlate well (within 95% in most breathing problems) with the "gold standard" of respiratory tests, ABG's (Arterial Blood Gasses). ABG's require drawing blood from an artery. ABG's are often preferred by doctors because they show not only the CO2 level, but also if the body is already using compensatory chemistry.

The issue of using oxygen frequently comes up when the insertion of a feeding tube is planned. My suggestion is that anyone who is planning a PEG insertion have their CO2 levels checked a few days before to determine the degree of risk from oxygen and a consult between the Pulmonologist and GI doctor inserting the tube concerning the need for NIV and use of oxygen versus changes to the NIV settings if sedation and pain meds slow respirations.

### ***NIV, Bi-Level, BiPAP, AVAP or a Ventilator?***

Any machine used to move air into the lungs is a ventilator. Some measure each breath given by the pressure, others by the volume, so a ventilator can be categorized as a pressure or a volume ventilator. That means diddly to an ALS patient having trouble breathing. What we care about is whether the air is delivered with a mask or requires a tube inserted into the airway. That categorizes ventilators into Non-Invasive Ventilation (NIV) with a mask, and Invasive Ventilation through an airway tube. Invasive ventilators don't have an abbreviation because for many years they were the only ventilators. They are commonly referred to simply as a vent.

#### ***NIV***

The first NIV machine was the Iron Lung which used negative air pressure to lift the rib cage and draw air in. The Iron Lung isn't used today but there are vest sized machines that still use negative pressure.

NIV machines using Positive Airway Pressure (PAP) were developed in the 1980s to help people with sleep apnea, a condition where the airway collapses during sleep causing the person to stop breathing repeatedly. By pushing air directly into the lungs through a mask worn over the nose, the machine keeps the airway inflated so it won't collapse. This is CPAP, Continuous Positive Airway Pressure. It is not the correct NIV for people with ALS because the constant pressure makes it difficult to exhale when abdominal muscles are weak. The result is a feeling of suffocation which is no improvement over not being able to inhale without the machine! If your doctor is recommending a CPAP machine, it is definitely time to see a Pulmonologist familiar with ALS.

#### ***BiPAP***

In the 1990s computer technology added a new dimension to NIV. Now the machine could push air in until a preset pressure was reached, then reduce the pressure to allow the person to exhale easily. Repeating this cycle made breathing more comfortable and suitable for people with

neuromuscular diseases who could not exhale against the higher pressure. This type of machine was sold by Respironics using the brand name BiPAP (Bi-Level Positive Airway Pressure). Like the patented brand names "Kleenex" and "Band-Aide", BiPAP has become the common name for all such products even though there are other brands of Bi-Level machines.

A typical Bi-Level machine's settings are limited to

- Spontaneous which only assists breaths initiated by the user.
- Timed triggers the user to take breaths a preset number of times a minutes.
- S/T combines the settings to allow the user to breathe at their own rate but triggers extra breaths as needed to assure a minimum number of breaths per minute. S/T is the correct setting for ALS patients.

Bi-Level machines continue to become more advanced and make important changes in respiratory care. Like CPAP, Bi-Level machines were developed for home use but are increasingly used in Emergency Departments, during procedures, and even in Intensive Care. Computer technology has added the ability to store details of use in newer machines which can help in adjusting settings for optimal use. This information can be brought up on the machine's viewer or using an SD memory card. Another change to newer Bi-Level machines is that nearly all now have alarms to alert caregivers to problems in delivering air. A change that is slowly coming is the addition of an internal battery. Without an internal battery, the machine has to be plugged in or be connected to an external battery to run. The use of external lithium battery packs has made them more portable than lugging around a car battery sized battery. A few Bi-Levels do have an internal battery that can power it for up to 5 hours.

The newest innovation in NIV is AVAP (Average Volume Assured Pressure Support) In the past, Bi-Level machines relied entirely on pressure to deliver the amount of air per breath. Volume monitoring was previously only available on Vents. AVAPS continuously detects the volume of air in each breath, averages it, and adjusts the amount of pressure gradually to assure the patient receives the right volume of air. Because the changes are adjusted whenever needed and are gradually applied, they aren't noticed by the user. Air leaks are adjusted for as well as changing masks and altitude changes. Like some other Bi-Levels, it can be set to gradually ramp up to its settings when you first put it on. All of this adds up to significantly more comfort as well as better control of CO<sub>2</sub> levels.

### *Invasive Ventilators*

There are several critical distinctions between NIV and an Invasive Ventilator:

- NIV requires only a mask, mouthpiece, or nasal plugs which can be put on or taken off as needed.
- Invasive ventilation requires either an endotracheal tube placed through the mouth or nose and down past the vocal cords to the trachea. This is an emergency measure. If it becomes apparent that the patient will need invasive ventilation for a prolonged time, a tracheostomy will be done to open a hole in the neck to place a tracheostomy tube directly into the trachea. A trach tube is far, far more comfortable than an endotracheal tube! For those who have decided to have a tracheostomy when Bi-Level ventilation becomes insufficient, that alone is good reason to do the tracheostomy before it is an emergency situation.
- NIV is not considered "Life Support Equipment." An Invasive Ventilator can be set to completely take over breathing. Bi-Levels can only assist breathing.

- The progression of ALS continues to weaken the muscles needed to breathe. A person on NIV will need to increase the hours per day of NIV use. NIV will reach the limit of its ability to sustain oxygenation and therefore life, so when 24-hour use is necessary, the decision on whether to change to invasive ventilation will need to be made.
- An Invasive Ventilator has more options in settings, including delivering higher pressures than Bi-Levels are designed to give. This makes it more effective and often necessary in severe pneumonia or blood clots in the lungs.
- Because NIV only pushes air in until the set pressure is reached, it won't deliver enough air when the lungs are filling up with congestion. It will reach the set pressure faster and switch to exhale, leaving you dangerously under-oxygenated, struggling for more air, and exhausting your breathing muscles. A trip to the ER and facing whether to go on invasive ventilation with a trach is next.
- ALS patients often reach the point of not being able to cough effectively. If a Cough Assist machine isn't effective and suctioning is needed by patients using NIV, the tube has to be passed through the nose or mouth. With invasive ventilation, secretions can be removed by passing a suction tube through the trach tube to remove secretions.
- The presence of the tracheostomy tube increases the risk of lung infections because it bypasses some normal protective barriers. Use of NIV also creates increased risk for lung infections and sinus infections as well as pressure sores from the mask or nasal pillows and the discomfort of air filling the stomach.

### *Hybrid Ventilators*

A growing trend is to use a hybrid ventilator such as the Trilogy which can be set for Bi-Level ventilation with a mask or mouth held tube, or for use with invasive ventilation. This option makes the transition to invasive ventilation easier if it becomes necessary. Medicare has ruled that if you use a Trilogy or other invasive ventilator for non-invasive ventilation, your co-pay will be higher than it would be for a Bi-Level machine.

One final comparison between NIV and invasive ventilation is the warning that a invasively ventilated person requires 24/7 care. That is true and yet misleading. How many people who are on Bi-Level ventilation during the day, whether part time or full time, are safe at home alone anyway? Arm weakness is usually a big part of the picture at this point so putting on or adjusting the headgear requires assistance as does using a cough assist. Power outages, equipment malfunction, toileting, choking, mosquito attacks, fire, etc. all require assistance 24/7 regardless of the type of respiratory equipment needed.

The care required can be provided by anyone given ten minutes of instruction on how to suction and the opportunity to practice doing so. A little info on how to troubleshoot a beeping vent and using an ambu bag and they are all set. No licensed nursing care needed.

The caregiver doesn't need to be in the room or even in the house. As long as they are close enough to get back in a minute or two and a reliable method for the PALS to summon them has been set up, they can have the freedom to spend the afternoon outside or at a neighbor's watching football.

<http://www.ventusers.org/edu/HomeVentGuide.pdf> is an excellent resource on all the types of ventilation devices. It shows those available in different parts of the world and the settings, size, weight, power source, and alarms on each type and brand.

## **BiPAP, AVAP**

### ***What is BiPAP, AVAP used for?***

BiPAP, AVAP (and their predecessor, CPAP) are most commonly used for people with sleep apnea or with hypopnea. Both of these can occur in ALS.

#### *Obstructive Sleep Apnea (OSA)*

The more commonly seen and therefore well-known apnea is obstructive. The muscles of the soft palate and throat weaken and the upper airway is narrowed or even collapsed when the muscles relax as the patient falls asleep. That blocks the airway and the patient wakes up repeatedly, often not aware that the reason he is awakening is because he quit breathing. Some people find they feel startled or even panicky with these awakenings, but other are so tired and groggy from lack of sleep they may not even be aware they are waking up over and over. Snoring generally occurs, but it is possible to have apnea and not snore at all. Similarly, obstructive sleep apnea is often associated with obesity however there are apneics who are of normal or low weight.

#### *Central Apnea*

Another type of apnea that can occur is "central apnea" in which the little nerve center in the brain stem that is supposed to regulate when we take a breath and how long we hold it, fails to work properly when we fall asleep. This can be a problem directly with the nerve center or with the motor nerves that carry the message to the muscles used in breathing. Even though we think of breathing as something that is continually on autopilot, sleep seems to somehow interfere with the autopilot mechanism and apnea occurs. All it takes is a little stimulation to get breathing going again -- for babies with sleep apnea, just jiggling the crib is often all it takes. For adults, a spousal elbow in the ribs usually does it! BiPAP gives a breath and that breath is either enough stimulation to get you to breathe again or is enough to tide you over until you do breathe again.

#### *Hypopnea*

A third possibility is that the muscles of respiration are weak and during sleep when breathing is normally shallower, breathing becomes too shallow. The patient doesn't stop breathing but breathing is inadequate. BiPAP can be used to increase the volume of air taken in without increasing muscle effort. This is the most common breathing problem for ALS patients.

### ***How does this apply to ALS?***

All three of the problems discussed above can occur in ALS. Bulbar weakness (weakness of the muscles controlled by the cranial nerves that arise from the bulb-shaped medulla portion of the brainstem) causes swallowing and speech problems and may also allow the collapse of the upper airway during sleep when muscles relax. This results in obstructive apnea. If ALS affects the motor pathways the brain stem uses to send the impulses to trigger breathing, central apnea can occur. And of course, ALS can affect the muscles used in breathing and cause hypopnea. Hypopnea is probably the most common problem in ALS but it certainly is possible for a combination of these problems to occur in ALS.

Whether it is a form of apnea or hypopnea, the patient does not get good quality sleep. The quality of sleep is not just dependent on the total number of hours, but also how that time is broken up by awakenings. In order to feel rested, it is necessary to get blocks of sleep that last at least 90 minutes -- that is when REM sleep occurs and REM sleep is apparently the stage of sleep where the brain has a chance to "recharge" itself. Without this good quality sleep, the patient becomes increasingly tired, has trouble staying awake during the day (yet has apnea and awakens if he falls asleep), finds it harder think clearly, concentrate, remember. Depression is very common. In addition to the mental effects, the lack of sleep begins to affect physical health too.

### *CPAP or BiPAP?*

CPAP isn't bad for people with neuromuscular weakness of the respiratory muscles in the sense that it is dangerous, it just doesn't work for them. CPAP is designed to give Continuous Positive Airway Pressure to keep the airway from collapsing on exhalation. In effect, it keeps the lungs partially inflated to the same amount during inhalation and exhalation. That makes it more difficult to exhale and that is why it is difficult for anyone to adjust to. For neuromuscular patients, the muscle weakness makes it even more difficult to exhale against the pressure. BiPAP/AVAP have separate pressure settings for inhaling (higher) and exhaling (lower). The lower pressure during exhalation makes easy so it is the correct choice for ALS patients.

### *How do I know if I would benefit from BiPAP/AVAP?*

Like everything else with ALS, the onset of breathing problems can vary from patient to patient. Commonly the person with ALS finds he gets a little short of breath when lying on his back. About the same time, it is likely he will notice that he becomes short of breath easily -- with exertion especially, but also after eating, when overly tired, or just in a stuffy room.

This gradually worsens and lying flat, whether on his back or side, becomes a problem. It becomes necessary to use several pillows or even begin sleeping sitting up or in a reclining chair. The human body was designed to breathe best when upright. Lying down makes it harder for the rib cage to move. Unfortunately, the human body was also designed to sleep lying down. Sleeping sitting up is not comfortable for anyone and it is really unhealthy for someone with ALS. If you sleep in a sitting position and then spend your entire day sitting, you will develop bed sores, more appropriately called pressure sores, on your tail bone and other bony points on your backside. This is not a possibility, it is an inevitable result if you spend 24 hours a day on your butt! People with strokes or spinal cord injuries may be unaware that they are developing a pressure sore. You won't be unaware because you haven't lost feeling, and pressure sores HURT! Also, constant sitting, even with your feet elevated, will not allow good circulation to your legs and swelling will be another problem. (See info on "Swelling of Feet and Legs")

Using a BiPAP/AVAP will allow you the wonderful and necessary pleasure of lying down, stretching out, turning on your side, and sleeping soundly.

A less obvious sign of hypopnea (shallow breathing) may be noticed before actually feeling short of breath. Even in healthy people, breathing is shallower when we sleep. For people with ALS, that little extra drop in volume can mean trouble since they are breathing shallowly to start with. When hypopnea occurs, you notice that you are more and more often waking up in the morning with a headache that goes away when you get up. The reason for this is that shallow breathing begins to affect the ability to "blow off" carbon dioxide. The excess CO<sub>2</sub> causes blood vessels in the brain to dilate which in turn leads to a headache. The headache goes away when you

wake up and start moving around because you breathe more deeply when awake and blow off the CO<sub>2</sub>.

We often associate that type of headache with "sleeping wrong" and tight muscles in the neck. Getting up also eases that muscle strain so differentiating between the two types of headaches is blurred. Try this: the next time you awaken with a headache, stay put. Don't change position, just begin taking deeper breaths. If the headache eases it is due to hypopnea and BiPAP/AVAP will help. Untreated, the hypopnea will get worse and you will begin to wake up more and more tired and groggy. You may begin to feel confused and disoriented in the morning and tired all day.

Although hypopnea is the more common problem with neuromuscular diseases, apnea can also occur. It can be really obvious that something is wrong. You find yourself drifting off to an exhausted sleep only to be jolted awake by a wave of feeling that includes a galloping, thudding heartbeat, weakness, dizziness, and a generally panicked feeling. This happens over and over until you finally are so exhausted you sleep through it. It can also be much more subtle. You wake up frequently and may or may not remember doing so. Either way, in the morning you know you just didn't get enough sleep. You are tired all day, doze off frequently, have headaches, feel half sick from exhaustion, have a poor appetite, lose weight dramatically, can't think or remember as well as you did, are emotionally on thin ice, and feel an undercurrent of constant anxiety. In short, you have all the problems associated with sleep deprivation even though you may seem to spend hours in bed and hours dozing in your chair.

An overlooked part of respiratory problems is the tremendous number of calories burned by the increased effort of breathing. Swallowing problems may not be present yet but the weight loss is fast and out of proportion to how much you eat. After a lifetime of battling the bulge, this is me before and after beginning BiPAP/AVAP.



If you can't breathe well enough lying down to get a good night's sleep, are sleeping sitting up, having morning headaches, or find yourself waking up frequently, or just feel like you aren't getting enough sleep, it is a pretty clear indication you have reached the point where BiPAP/AVAP could really improve your quality of life. I sleep better when I am wearing it, but that is only part of the benefit. Since I began using BiPAP at night, my days have improved drastically. Not only do I have less trouble breathing during the day, I eat better, choke less, feel stronger -- mentally, physically, and emotionally, have more stamina, am not cold all the time, my heart is not pounding along at 100 beats per minute, and, in general, feel like I took a giant step back from death's door.



## ***How to get BiPAP/AVAP***

You have to have a physician's order (prescription) to get a BiPAP/AVAP machine even if you do not plan to seek insurance reimbursement so the first step is:

### *Finding a Doctor*

Unfortunately, finding someone who is "cross trained" to understand both the effects of neuromuscular disease on respirations and the respiratory assistance machines available can be hard. The neurologist knows neuromuscular disease but diddly about the machines. The pulmonologist (physician specializing in respiratory problems) knows the machines but not neuromuscular disease.

Start with your neurologist. A neurologist who works with ALS patients should certainly be familiar with NIV. He may handle your respiratory care himself but may prefer to refer his patients to a pulmonologist for respiratory care at that point. I would be reasonably comfortable with either of them. In our screwed up health care delivery system someone might require that you see a sleep specialist. The sleep guys are used to sleep apneatics who have good lung power and may be unfamiliar with the problems of the neuromuscular patient. If I were sent to a sleep specialist I would most definitely ask "How many ALS patients on BiPAP/AVAP have you cared for?" and if he could not demonstrate a very good understanding of ALS, I would be looking elsewhere.

The doctor who handles your respiratory care is going to be a very important person in your care. Patients with ALS who do not want to go on a ventilator most often die of respiratory failure so this doctor will be the one who sees you through to the end or sees you through many years on a ventilator. You may still see other doctors, but odds are that this is the one who will be there directing most of your care. You need someone who is comfortable not only in deciding what pressure settings you need and dealing with any respiratory infection, but also in listening, explaining and helping you make decisions about things like tube feedings, code status, and whether or not to switch to full ventilation when BiPAP/AVAP is no longer sufficient and perhaps when to discontinue the ventilator. At some point, he or she will likely be the physician you depend on to make the end as easy and dignified as possible.

### *Undergoing Testing*

There are several tests doctors can do to monitor how much ALS is affecting breathing:

- A respiratory function test involves huffing and puffing into a machine to see how much lung capacity you have. The FVC (Forced Vital Capacity) which is basically how deep a breath you can take is one of the most important findings of the test. At about 70% of normal, you begin to notice problems, less than 60% is considered moderate, less than 40% is severe. People with neuromuscular diseases should have the test done while they are lying down as that is when their breathing is the most impaired. A respiratory function test/FVC does not show whether you have sleep apnea or if hypopnea is even worse when you fall asleep, however.
- Another helpful test is a simple test of blood oxygen level. This is reported as Oxygen Saturation (O2 Sat) percentage. An O2 Sat of 96% means that 96% of your red blood cells are carrying a full load of oxygen (are saturated) like they are supposed to be. Normal is 90% or better (usually high 90's). O2 Sat monitoring can be done with a simple little device you clip or tape on your finger tip. No needles! For us, the important thing is to wear the



device while lying down and sleeping. A spot check in the middle of the day will no doubt show excellent levels when breathing problems first begin because our breathing is at its worst when lying down and/or sleeping. To accurately test you, your doctor can arrange for you to use an O2 Sat monitor at home overnight. You just put the device on your finger at bedtime and wear it all night. It monitors oxygen saturation and heart rate and records it. The next day you return it, the results are retrieved from its memory and reported to your doctor. If you are breathing too shallowly (hypopnea) your O2 Sat should drop. (CO2 levels would be more accurate in spotting hypopnea but as discussed below, they are not as easily done accurately.) If you have apnea, your O2 Sat will drop briefly but dramatically at repeated intervals.

These two tests along with a diagnosis of a neuromuscular disease such as ALS and a description of the problems you are having should be sufficient to get a prescription for BiPAP/AVAP and insurance reimbursement for it. Unfortunately, sometimes it gets more complicated. Physicians and/or insurance companies may want more diagnostic evidence and want other tests done.

- ABG's (Arterial Blood Gases) require drawing blood from an artery (usually in the wrist -- an "uncomfortable" procedure). ABG's can tell a doctor a lot more about your respiratory status than just oxygen levels. For example, they tell the CO2 levels, can help determine if your breathing problems are due to lung problems or other problems such as liver or kidney disease and even how well your body is able to compensate for the problem. But ABG's are NOT always necessary.

Although ABG's are a perfect way of checking CO2 levels, they are a total waste of time for the early stages of breathing problems in ALS if they are done when you are wide awake and breathing normally! When breathing problems become severe CO2 levels will be high even when you are awake, but not early on when you first could begin benefiting from NIV. DO NOT allow them to draw ABG's during a daytime appointment (unless you have some other lung disease too) as a first step in determining if you need NIV. You have to be in pretty bad shape to have a high CO2 while you are wide awake and being stuck in an artery with a big needle! Doctors tend to be irritatingly fond of this test. INSIST that the doctor explain what he needs to know from the ABG's that he can't figure out simply by knowing that you have ALS and overnight O2 Sat monitoring.

- Some physicians and/or insurance companies will want you to have a sleep study done to verify the need for NIV. This expensive test involves spending a night in a sleep lab trying to sleep while you are all wired up with monitoring devices. The information will tell the doctor whether you have sleep apnea and help determine what kind of pressure settings your machine will need. A sleep study is necessary to get a diagnosis of Sleep Apnea, but for an ALS patient that should not be necessary. The use of BiPAP/AVAP for the breathing problems accompanying ALS is well documented and standard care. Simply having a diagnosis of ALS and evidence of the onset of breathing problems should be sufficient to justify ordering Bi-PAP. A secondary diagnosis of sleep apnea or hypopnea by a sleep lab should not be needed. But, depending on your insurer, you may have to jump through their hoops. Again, your doctor may believe that the insurance company will reject a claim for BiPAP/AVAP without this bit of supportive evidence but I would ask him to file with all the other info he has on you first and would not agree to the test unless the insurance company insisted on it.

If you are required to have a sleep study, be aware that ABG's are commonly done with them. Again it is important that the blood be drawn while you are asleep -- or as soon after awakening as possible. (No one is going to sleep through an arterial blood draw!) Ideally, the blood should be drawn first thing upon awakening. I would refuse to have the test done if it was not done at least before I was out of bed. I have heard of instances where it was done after the patient was up to the bathroom, dressed, and ready to go home! No wonder the ABG's did not support her need for NIV! You always have a right to refuse a test or medication, and in this case not only is the test useless but it will work against you in getting reimbursement for NIV!!!

### ***How much does BiPAP/AVAP cost?***

A BiPAP/AVAP machine can cost anywhere from \$950 to as much as \$6,500 depending on the features it has. They are covered by most medical insurances. Although it is possible to purchase your own machine, the most common thing is to rent one from a respiratory care company in such a way that you eventually own the machine. I have found that renting a machine is well covered but sorting through the finances is a headache. No one seems able to explain it fully. Therapists are seldom well versed in the billing process and billing people only seem to know what your current statement shows. Anyway, the common way of renting NIV machines is called Capped Rental.

Medicare and most private insurance companies do a capped rental on BiPAP/AVAP machines. As with most durable medical equipment, they will pay 80 percent of the rental fee up for 13 months. Once they have paid that much in rental, the machine is considered paid for and is no longer billed.

In the past, once the 13 months were up you were given the option of owning the machine but not receiving any further visits, maintenance, or repairs from the supplier, OR agreeing that the machine would continue to belong to the supplier and be returned to them if you no longer needed it or changed supplier. With the second option the supplier had a service contract to provide visits by a Respiratory Therapist to check the machine or adjust the settings or help you find the mask that works best for you. Maintenance, repairs and replacement were the responsibility of the supplier at no charge to you.

In 2006 Medicare dropped the second option by placing BiPAP/AVAP in the category of medical equipment that does not need frequent or substantial servicing. Now, after 13 months the machine belongs to you and the supplier has no service contract and provides no further services.

By 2016 machines such as the Trilogy, which can provide BiPAP/AVAP and, when needed, switch to invasive ventilator settings were becoming common. These machines are considerably more expensive, so Medicare changed their reimbursement to just two categories;

- Respiratory support provided through a face mask or other non-invasive method.
- Respiratory support provided invasively through a tracheostomy tube.

Rather than allowing the Trilogy to be filed as a BiPAP/AVAP machine, Medicare ruled that it must be filed as a full, invasive vent regardless of how it is used. The end result is that if you use a Trilogy as a BiPAP/AVAP, your co-pay will be higher than it would be for a NIV machine.

A BiPAP/AVAP machine is expected to last for at least 5 years and Medicare will pay for any needed repairs for 13 months, but after that, the routine maintenance is up to you, as is dealing with mask problems and setting changes.

Replacement of masks, tubing, etc. is 80% covered by Medicare on their time schedule: 90 days on masks, 2 pairs of nasal pillows and new tubing per month, 180 days on headgear.

There are no doubt variations from one insurance company to another, but it is likely that insurers will follow Medicare's lead and end their service contracts with BiPAP/AVAP suppliers.

### ***My First Night on BiPAP***

I'd had a very slow progression of ALS (first definite problems noted 9 years before I began noticing some breathing problems). At first it was just feeling a little short of breath when lying on my back. With time that got more definite. I began waking up with headaches occasionally but not consistently. I saw a pulmonologist (physician specializing in respiratory problems) and had pulmonary function tests done. My function was at 70%, still good.

After time passed and my weakness progressed, I reached the point where I had to sleep on my side and even that became uncomfortable at times. I got a hospital bed so I could sleep with the head of the bed up and that helped for a while.

Then I started experiencing repeated awakenings with an awful rushing feeling starting in my chest and spreading. My heart would be racing and pounding. It was not too bad during the day though there would be stretches of an hour or two when I was uncomfortable even though sitting up. But nights were awful. I would eventually fall into an exhausted sleep by turning on my side while having the head of the bed up 45 degrees, but you can imagine what my back and shoulder felt like after a couple of hours. It took hours to fall asleep and I was waking my husband up so many times a night to be repositioned, neither of us were getting enough sleep.

I went to see my pulmonary doctor again and he recommended a "poor man's" sleep study. Rather than have me come into the Sleep Center for an overnight full monitoring session, he sent me home with an O2 Sat. monitor. That is a little machine that has a fingertip sensor you tape on to detect what your oxygen level is. It also takes your pulse. I wore it overnight (a fairly good night) and we found that my O2 dropped to 65 % at times. Normal is over 90%. My pulse was humming along at 90-110 even in my sleep. Normal is 60-90. I was promptly visited by a home respiratory care agency and they set me up with the Bi-PAP machine. (No attempt was made to define whether my problem was apnea or hypopnea, nor were ABG's needed. The insurance accepted my claim on the first try and didn't ask for any further proof that I needed BiPAP/AVAP .)

The machine itself is small, about the size of a 4 slice toaster. It has a hose that attaches to a soft, flexible plastic mask that fits over your nose. (Doesn't cover your mouth.) There are Velcro straps on the mask that fit around your head so the mask fits snugly and air doesn't leak out. When turned on, the machine blows air into the mask. It doesn't use oxygen, just regular room air but it can use oxygen if you need it.

Now the fun part! The therapist selected a mask she thought would be the best size for me and went about strapping it on. Imagine being short of breath to start with -- not just a momentary event but a couple of months of feeling miserable in stuffy rooms and suffocated by lying flat -- then imagine having someone put a plastic mask over your nose. Instant panic! Being the kind of person who never learned to swim because I was terrified of water up my nose and dunking my head didn't help at all!

"Where is the Valium??" I whined as I sat forward in my chair and gasped for air.

I convinced myself I could breathe through my mouth just fine and relaxed after a moment and let her work with the straps. I told her I thought a lot of my anxiety came from the fact that my arms are so weak and I knew I wouldn't be able to just grab the mask and pull it off by myself. "A control issue," as they say. She said I needed to breathe through my nose when the machine was on or the air pressure was lost through my mouth.

"OK," says I, gamely.

She turned the machine on . . . Ever ride in a car at 80 miles an hour with your head out the window? All that wind and you can't get a breath of air because it is moving so fast! The mask was not snug and air was whooshing out and I opened my mouth to take a breath and my ears popped from the change in pressure. I found some arm strength I didn't know I had and off it came!!!

She assured me it would feel fine once the mask was snug and I braced myself to try again. Mask on, air blowing wildly in my face as she worked with the straps and I gasped through my mouth. "There," she said. "Breathe through your nose!" I tried. Snorted. Tried again. I found that the pressure wasn't noticeable at all as far as taking a breath was concerned. It was just as easy to breathe with the mask on. I sat there, trying hard to stay halfway relaxed as she adjusted the straps.

Just as I was thinking I might be able to tolerate this, I had to speak to answer her when she asked if there were any leaks or if the straps were uncomfortably tight. When I opened my mouth the air rushed out and my ears felt the pressure change. What a weird sensation! It was hard to talk around the pumps of breath from the machine. Yuck!!!! I wanted out! She encouraged me to keep breathing through my mouth.

It took me a half a minute to settle down again and just breathe, but when I did I could feel a tremendous difference. WOW! It worked! I was rewarded with the first full breaths I'd had in months! That tight, achy, tiredness in my chest that I had been living with was eased. That was all the encouragement I needed to keep going. She decided I needed a different size mask as there were still some small leaking areas and it seemed to be putting a lot of pressure on the bridge of my nose. We swapped masks and tried again. Again the process of getting it on was icky, but once it was halfway snug it felt not just tolerable, but good. I didn't really want to take it off when she left, but neither did I want to be home alone with this weird contraption on me!

My husband had not been able to be home during all this, so when we went to bed that night, he had to get the thing on me without instruction. It is quite simple but he was not prepared for me to get all panicky while it was loose and the air blowing in my face. (The therapist had recommended having the machine on while putting it on.) We managed though and I was soon breathing happily if a little tensely as I experimented with such death defying maneuvers as tilting my head and swallowing. I stayed sitting up for a long time but just couldn't fall asleep and finally felt secure enough to try putting the head of the bed down. No problems! It required a little adjusting of the fit as it wanted to leak a little air, but that was easily fixed.

But I still couldn't get to sleep. I sleep best on my side and decided to give it a try. I woke up my husband, told him what I wanted (getting better with the talking part) and he turned me. Wiggled the mask a little to seal the new leaks and I was comfy and snoozing shortly.

A couple of hours later I awoke in a panic and yelled for him to "Get it off!" It took a little doing to get it undone in the dark. I decided I'd had enough fun for one night and left it off. But now breathing was uncomfortable. I had him reposition me a couple of times, put the head of the bed back up, and soon decided I really wanted the mask back on. Long suffering hubby made irritated noises but put it back on. This time I had him leave the machine turned off until the mask was in place and that worked better for me.

Finally convinced that I was in control -- it wasn't forcing breaths on me or fighting my breathing at all, I curled up on my side and slept for 4 hours straight -- except for one little wake-up by our kitty who had decided it was time to check out what idiocy I was up to now with that thing on my face!

She sniffed carefully at it, tickling me with her whiskers, sat and stared for a bit, sniffed again, then turned away. "Whatever gets you through the night," she shrugged as she wandered off.

I agreed.

I didn't realize how pervasive lousy breathing is until after a week or two of using the machine at night and for a rest period during the day or evening. I had been feeling so bad in so many ways and never associated it with the breathing problem! I had been tired, weaker than ever, totally lacking in stamina. I simply sat and played solitaire on the computer all day. I experienced periods of dizziness, feeling faint, feeling nauseated. I had no appetite and felt just stuffed after a few bites (a real change for me!!) so I was losing weight. I was choking more easily, and if I did eat, I had abdominal cramps and nausea immediately afterward. I was miserably cold most of the time. It wasn't that the air around me felt cold, it was like my bones were icicles and the cold was seeping outward. I knew my heartbeat was too fast and suspect that my blood pressure was very low. Emotionally I was outwardly OK but I could feel a constant state of anxiety that somehow existed side by side with a sort of mental sluggishness and lack of initiative. Anything that seemed to make it harder to breathe -- a stuffy room, a hug, lying down, sent me into a momentary panic. Making love resulted in panicky gasping for air. Bed time was no relief. In fact, it was the worst because I knew I was going to be jolted awake over and over.

Every one of those problems started to improve immediately with the Bi-Pap. That isn't intended as a testimonial ("Step right up here folks! Get your Dr. John's Famous Elixir! Cures apoplexy, nervous conditions, gout, and sweetens your breath!") but simply to show you how breathing problems sneak into everything your body does. I had no idea that most of my problems were related to my breathing. I can't promise you that you will have this kind of results, but I do think you will find that overall you feel a lot better if you start using Bi-PAP. (And no, I do not own stock in Respironics Co.!! Wish I did!) Having to wear a stupid looking mask every night, putzing around to get comfortable and get rid of annoying little leaks, the annoying cold stream of exhaled air, dealing with a lumpy head of hair and facial crease marks from the mask every morning is not fun, but it is so much more tolerable than living without BiPAP/AVAP.

# **Ventilators and Trachs**

## ***The Vent Decision***

If you are evaluating the pro's and con's of going on a ventilator when the time comes, there are big questions you need answers to. How will affect my quality of life and that of my family? What help will I need at home? What will it cost for the equipment? If living at home becomes a problem, how far is a nursing home that accepts ventilator patients? It is hard to answer these questions because insurance coverage varies, the availability and willingness of family and friends to help varies, and each individual's adaptability to advancing ALS is different.

Breathing is not a problem if you opt for a vent and trach. Swallowing is not a nutritional problem with a feeding tube. That pretty much takes care of the actual life threatening problems of ALS!

A vent is not for everyone. Those with rapid progression and loss of nearly all movement will face being locked in (mind is working but with no way to communicate even yes/no) fairly soon. In all of the United States, a patient can request to be taken off the vent and allowed to die at any time, and the process will be done under heavy sedation and other meds for a peaceful death. Learn about your country's laws regarding your right to discontinue treatment, including a ventilator, before deciding.

The hard part is deciding "when". If the patient waits too long, he or she can be locked in with no way to say turn off the vent. Patients can live for years in this state. Most people consider that an extremely undesirable way to live. Family is stressed terribly. No one should go on a vent without deciding when they want the vent turned off and telling their family and doctors that decision. Giving medical power of attorney to someone they believe will carry out that decision is critical. Often the best person for that job is not an emotionally torn family member.

Going to a vent is not an easy decision. If you have read "Safe Harbor" you know that I have found vent life to be positive and I tend to be pro-vent. It is horrifying to me that only 5% of ALS patients in the United States opt for a vent while 50% of ALS patients in Japan do. But that is a topic about our pathetically bad health care system...

I definitely recognize that I have had all the breaks in the situation; a husband who was able to take early retirement to care for me, supportive family even though only one family member lives here and the rest are 800+ miles away. A couple of friends have pitched in to help so we don't have the expense of paid care. I am a couch potato by nature and can keep occupied and happy. Most of all, my ALS has been very slowly (but consistently) progressive making the decision to vent worthwhile. In spite of all that, I have tried to present the information here in an unbiased way as I can. I hope you find the information useful as you try to decide what to do.

## ***When a Vent and Trach is Better Than Non-Invasive Ventilation***

When neuromuscular disease weakens the muscles used for breathing, many people benefit greatly from non-invasive ventilation (NIV), which can add years of breathing support. But when a ventilator and tracheostomy are suggested for better breathing, some people see BiPAP/AVAP as "enough" and a vent and trach as "too much." When a person's overall quality of life is no longer acceptable, that's certainly a valid choice. But a trach and vent shouldn't be ruled out if quality of life problems are due in large part to respiratory problems.

## *Why Switch from BiPAP/AVAP?*

BiPAP/AVAP assist breathing through face masks, nasal plugs or tubes to “sip” air, without the need for surgery. So why would anyone even consider going to a trach tube? BiPAP/AVAP can prove inadequate for a variety of reasons:

1. Facial features such as a crooked nose or a deviated septum can make finding a mask that doesn't leak or breathing entirely through the nose difficult and very frustrating. Although most masks are adjustable and therefore called "customized", truly custom made (molded) masks are now available. Most insurance limits the number of masks per year making finding the right one expensive, and insurance may not cover a custom made one. Adding to the problem is that the facial contours change significantly with weight loss or gain and development of facial weakness.
2. BiPAP/AVAP may aggravate sinus problems and lead to chronic sinus infection.
3. For some people, BiPAP/AVAP pushes air into the stomach and causes very uncomfortable abdominal distension.
4. Some find anything on the face claustrophobic.
5. Facial weakness reduces necessary jaw closure and ability to use a mouthpiece or mask.
6. It can take months to find the right settings and mask or device and get used to NIV, so if knowledgeable support or strong motivation is lacking, BiPAP/AVAP probably won't work out.
7. When BiPAP/AVAP use extends into daytime use, going out becomes more difficult because of equipment needs, although BiPAP/AVAP units are being changed to be more portable in terms of size and portable power supply. Wearing the mask in public is unacceptable to most users because of the headgear. Nasal plugs still require headgear -- they aren't the simple tube worn by oxygen users. BiPAP/AVAP headgear definitely looks more "Star Wars" bizarre than "Top Gun" cool. Although a sip tube is much better in that respect, its use is limited because the patient must be able to form a seal around the tube and suck strongly enough to trigger the machine to deliver a breath.
8. Ventilators have an alarm system which a BiPAP/AVAP machine may not have.
9. Using an invasive ventilator for BiPAP/AVAP greatly eases the transition to full ventilation if that is being considered. Familiarity with the machine goes a long way even though the settings are different.
10. The most common reason for switching, however, is that after successfully using BiPAP/AVAP for some time, a person's breathing muscles weaken further. Several symptoms show that this is occurring, such as:
  - Frightening spells of suffocating or choking congestion caused by thick mucus and a poor cough. Breath stacking, manual cough assistance or a CoughAssist device work very well for some, but others find these methods minimally effective. The inability to clear congestion is exhausting, frustrating, and dangerous. A trach solves this problem.
  - The hours the person needs to use BiPAP/AVAP increase from overnight to most of the day.
  - Because BiPAP/AVAP can only assist breathing, not entirely breathe for the user, as a person's breathing capacity deteriorates, he or she slides back into the fatigue, poor appetite, and anxiety of pre-BiPAP/AVAP days.

- Pneumonia or a simple chest cold results in a respiratory crisis. The additional settings available on an invasive ventilator may be needed and the ability to remove secretions is critical. At this point, a trach is generally more effective than CoughAssist.

#### *Why the Resistance to a Breathing Upgrade?*

One problem is, as so often the case, cost. Because an invasive ventilator is capable of more than delivering more than a Bi-level Ventilator, it is more expensive. For this reason, most people are limited to BiPAP/AVAP until the switch to full ventilation is required. Cost is certainly a factor but it is hard to discuss cost because insurance coverage varies drastically. Some people have complete coverage, others have a sizable copay. The best I can do is give you my cost. (Remember, this is my cost and every equipment supplier and insurance is different!) Go to Cost of Vent Equipment.

It is possible to purchase your own vent but you will not receive any of the services provided along with vent rental. When you rent, a Respiratory Therapist visits frequently at first, then less often. The therapist does no patient care. The therapists are there to check the equipment, not the patient. If I have any problems the therapist can intervene to work with the doctor to change settings or recommend other equipment. During visits, the therapist will run checks on the vents, suction machines, batteries, chargers or other equipment. They arrange for prompt replacement if needed. They keep records of all the machines so they are regularly exchanged and sent in for a full "tune up". They are on call 24/7 for equipment emergencies. Equipment is replaced at no charge if there is a problem with it. We order our supplies by phone through Apria as well and they are very prompt with most items.

Perhaps another source of resistance is all the emotional baggage the word ventilator carries, primarily the words "Life Support." Sadly those words always conjure up images of death bed scenes where life support is withdrawn. Yet the reality is that ventilator use is far more positive in the vast majority of cases. Premature babies, people having major surgery, accident victims, people with severe pneumonia all survive because of ventilators. And so can ALS patients. Granted the differences for ALS patients are major; permanent ventilation versus temporary, advancing disability versus recovery. But when the major obstacles to quality of life are the fatigue, loss of appetite, anxiety, depression, repeated bouts of life threatening congestion all caused by respiratory problems, a ventilator can restore quality of life at least for a while.

One of biggest reasons for resistance to venting is, sadly, negative comments and advice from doctors and nurses. The information you get from doctors and nurses is so out of touch with reality! Some of my favorites are:

- ✕ *"You won't have any quality of life being tied to a ventilator."* Well, the patient is the only one who can judge his quality of life and if it isn't satisfactory, he can have the vent removed. My quality of life improved drastically with the trach and vent. See quality of life studies, Page 63.
- ✕ *"You can never be left alone if you are on a vent."* True. But is it really possible or safe for you to be alone if you are at the point of needing a vent? No! A good call system is essential either way.
- ✕ *"You will need expensive round the clock nursing care."* Nonsense. Family and friends can do it. Managing vent maintenance requires an hour or so of training but anyone, including kids and probably chimpanzees, can suction, handle vent alarms, and use the ambu bag if necessary with just a few minutes of explanation and hands-on time.



- x *"The burden on your caregiver(s) will be heavy."* A significant increase in care needs is far more likely to be the result of the advancement of ALS weakness rather than going on a vent. The additional work for your caregiver(s) because of the trach and vent will be suctioning you (less often as time goes by) and daily cleaning of your trach area and, depending on the type of trach you have, cleaning the trach tube. After a period of adjustment, I think you will find your caregiver is actually less stressed, at least by the breathing aspect of your ALS. Congestion that now leaves you both wondering if you can cough it out or if you are going to die this time will be nipped in the bud with suctioning. Any choking spell can still be exhausting but the trach and vent will assure that your airway can't be blocked and you will continue to breathe throughout the choking spell.
- x *"Your house will look like an Intensive care unit."* Only if you want it to. Suction machines and other equipment are right at hand even in a cabinet or drawer. Trach supplies don't need to be sitting out anymore than your toothbrush and deodorant do. Being in a wheelchair required infinitely more expensive and highly visible changes to my home than going on a vent!
- x *"You can't taste or smell when you have a trach because the air isn't passing through the nose."* This falsehood is all the more irritating because it is a quote from Dr. John Bach, the guru of non-invasive ventilation. (Quoted in "MISSION POSSIBLE?: Converting to Non-invasive Ventilation By Jean Dobbs for New Mobility Magazine, May 1996.) This may be true for some trached people, but definitely not for all! My sense of taste is, unfortunately for my waistline, unimpaired. My sense of smell is limited only in that I cannot sniff well enough to smell something being waved under my nose. A scent that permeates the air such as food cooking, a vase of flowers, or something going bad in the fridge is very obvious to me!
- x *"Trachs are complicated, time-consuming, problem-prone, ugly, uncomfortable, expensive, prevent talking and eating."*

These comments deserve a separate page for disproving! See "Trach Care", Page 68.

## ***Quality of Life***

Breathing is not a problem if you opt for a vent and trach. Swallowing is not a nutritional problem with a feeding tube. That pretty much takes care of the actual life threatening problems of ALS!

What is left to consider are quality of life issues. Loss of speech is a hurdle that many people find very difficult to deal with emotionally. I haven't reached that point but suspect that the speech equipment available will work well for basic communication but can't give back the pleasure of a simple conversation. Anyway, loss of speech won't happen because of the trach itself (and the vent will give you the air volume for a louder voice), but because of the progression of the ALS. With Bulbar onset, speech has to be considered very temporary, as does swallowing.

As the ability to swallow worsens we face the "socially unacceptable" and therefore embarrassing problem of drooling. The good news is that it can generally be reduced to an annoyance rather than a major problem.

If all of that seems like something you could deal with, there is still the need to recognize that the ALS will continue to progress. Losing the use of your legs is a walk in the park (bad pun!) compared to losing the use of your arms. A wheelchair and lift system or Hoyer lift pretty much

solve the leg problem. Arm weakness will eventually require an eye tracking system to allow you to do anything with a computer and that seems critical in giving any outlet for an active mind.

Bottom line: All of these quality of life issues are going to happen whether or not you choose a trach and vent. The difference is that they and you will go on longer if you do. Do you and your family want the extended time under those conditions.? Can you accept that your progression might be so fast that the extended time is mostly after you have lost all function? Not wasted efforts but probably not nearly the quality time you had hoped for. Can your family accept the flip side of that, that you could live for years?

*What can a ventilator do for your quality of life?*

- ✓ Relieve shortness of breath. It will not just ease breathing difficulty, it will completely erase it.
- ✓ Alleviate the underlying feelings of anxiety and nervousness or even panic attacks that being short of breath causes. Those feelings are not mental or emotional indications that you aren't coping well with ALS. They are normal physiological reactions that poor oxygenation triggers in everyone. If you are drowning or suffocating your body wants you to rev up and fight and not just relax and say "What, me worry?" That reflex remains strong in ALS.
- ✓ Stop headaches caused by low oxygen.
- ✓ Allow you to eat or have tube feedings without your body rebelling against having to deal with shifting blood flow and already scarce oxygen to digestion.
- ✓ Improve your energy level so that you can enjoy your family, have a social life again, go places and enjoy things without total exhaustion that lingers for days.
- ✓ Make you look better with pink cheeks instead of gray and without the dorky looking head gear and mask of NIV.
- ✓ Allow weight gain because your calories can be stored rather than sent immediately to keep your struggling lung muscles going.
- ✓ Let you think more clearly -- and worry about something other than breathing!
- ✓ Spare you (and your family) from those terrifying sessions of congestion when you cough and cough but all your weak efforts only succeed in bringing the mucus to your upper airway and threatening to block it entirely. With a trach and vent, those awful and life threatening minutes are reduced to a quick, painless and routine to the point of monotonous suctioning.

*What are the things that a ventilator cannot do for you?*

- x Slow progression of ALS. With fast progression, a ventilator can relieve the breathing problems, but the ALS will continue to progress. If speech and swallowing haven't already been taken away, they will be, along with any remaining muscle function. The question becomes whether or not venting will give you quality of life if every thing else disintegrates rapidly. It must be said, however, that there certainly are people who find life worth living without the ability to speak or eat.
- x Relieve pain and overall discomfort. People with fast progression have the most pain, probably because the ligaments and tendons have little time to adapt to the changes in muscle tone. In slow progression that is less a problem and as an extra bonus the horrible muscle cramps and irritating fasciculations taper off as more and more muscles fade away.

- x Pneumonia is a common cause of death among ALS patients whether they are on a ventilator or not. If you have had more than one bout with pneumonia you will probably remain prone to it. If it was due to aspirating food or saliva into the lungs, a feeding tube and a cuffed trach will be more helpful in preventing pneumonia than the vent itself.
- x Improve other health problems. Poor health and chronic diseases may be helped by better oxygenation but they will still be there. Age is certainly a factor when considering a vent but age is more than a matter of numbers. If you were physically fit and in good health before the ALS and have a family history of long life, then a ventilator isn't a bad choice even if some people consider your numerical age a problem.

Quality of life is hard to anticipate or for anyone else to determine. We have all heard that people can deal with far more than they thought they could, but that ability only stretches so far. How far? Studies have shown that evaluations of a persons quality of life by doctors and other professionals are consistently lower than the person themselves rate it:

- A 1992 life satisfaction study by John Bach compared responses by 80 Duchenne muscular dystrophy ventilator users to responses by 273 MDA clinic directors. The clinic directors significantly underestimated the users' reported life satisfaction. The study concluded that patients who were perceived by physicians to have a poor quality of life were less likely to be offered assisted ventilation.
- In a 1992 study by Bach and Campagnolo of 395 ventilator-assisted people who'd had polio, 86 percent reported their lives were characterized by hope, value, freedom, and happiness. Forty-two percent of the ventilator users were employed and 39 percent were married. Yet again, the health professionals used for controls underestimated the life satisfaction of people using ventilators and overestimated the difficulties. It was concluded that physicians should be aware of their inability to accurately gauge the life satisfaction of individuals and should not use their perception of poor quality of life as a reason to deny ventilator assistance.
- In a 1994 life satisfaction study by Bach and Tilton, 42 ventilator-assisted quads and 45 autonomously breathing quads were compared to a control group of health care professionals. Both groups with spinal cord injury reported higher life satisfaction ratings than were predicted by the controls. Notably, the ratings for those who used ventilators were higher in some areas than for those who didn't -- the former were dissatisfied with sexual function only, whereas the latter were dissatisfied with their jobs, health, and sexual functioning. Vent users said life was friendlier, more interesting, more enjoyable, fuller and more hopeful than non-vent users.
- A 1992 study by Whiteneck, Charlifue and Frankel of people with spinal cord injury at least 20 years post- injury showed that those using ventilators rated their quality of life higher than those who didn't, and had a lower rate of suicide ideation.

*The above information was taken from "Independent Living and Quality of Life Among Persons Who Use Ventilators," an in-press book chapter by Margaret A. Nosek, Ph.D., and S. Ann Holmes, M.D. Both authors are ventilator users themselves.*

These studies are old, reflecting the time period when home ventilation was first possible. "Allowed" may be a better word. I haven't been able to find more up to date studies which is frustrating. Improvements in technology have made huge differences in quality of life on a ventilator. Vents have gone from the size of a large microwave with no battery power or portability

to the size of a large lunch box or a large laptop that can be placed on a wheelchair. Lithium battery packs provide about 6 hours of power and are small enough to carry more than one on the wheelchair. Computers have eye tracking capability that allow completely hands free use. Lifting equipment and slings have also improved greatly as have beds and mattresses, wheelchairs, and so much more that quality of life on a ventilator today should definitely be re-studied. Twenty-five years has made a big difference!

Each of us has a tipping point where quality of life goes from acceptable to unbearable. Where that point lies is different for each of us. Unfortunately, we can't foresee or accurately predict that point. Many people settle the issue by drawing a line in the sand early on and saying "No vent!". And that is right for some people. But that line is drawn in sand and sand can shift. Time can change technology, our circumstances, our view of our disabilities. Drawing that line can give resolve and emotional peace. Drawing that line can also lock us into a promise made not to vent that makes it hard to admit that we may be considering otherwise.

### *Emotional Considerations*

Life on a vent requires adaptability. Can you find things to occupy the hours? You will be able to be out of the house much better, but there will still be most days spent at home. A computer is a great source of entertainment, news, interaction with others if you are willing to work with one. By the time you need to decide about going on a vent, you will probably be very reliant on others for care and have a good idea of their ability, willingness, and tolerance for continuing to care for you. There is nothing more demoralizing than a main caregiver who is resentful, angry, or acting the martyr. Family and friends may have already deserted you. Good riddance to the lot of them. Your spouse, partner, child, or parent will most often be your primary caregiver and their health, availability, and suitability must be considered. Regardless of their willingness, they must have help. Someone must be shown how to suction you and commit to at least a few hours a week at a scheduled time. Ideally, you will have more than one person for this so that your primary caregiver can get out to get groceries, keep appointments, etc. People uncomfortable with taking care of you can commit to things like lawn mowing, emergency shopping,

A trach, vent, and feeding tube can keep you alive indefinitely. At some point (assuming pneumonia, heart attack, stroke, flu, or killer bees don't get you first) you may decide it is time to explore the great beyond. Whether you have BiPAP/AVAP or trach and vent, dying will be similar, just at a different pace. (With a Bi-level Ventilator or invasive ventilator you can choose when, without them you have to wait until the process of dying begins naturally.) You will be given sedation to minimize anxiety and morphine to reduce the feeling of "air hunger" (polite medical term for suffocation) by dampening respiratory drive. If you aren't using any type of breathing machine, you will continue these comfort measures for days, weeks, possibly months. Doses will be increased as needed to keep you comfortable. Eventually, you will slip into unconsciousness and die in a few days.

If you have ordered that the breathing machine be discontinued, the process will be faster and the unconsciousness hurried by heavier sedation and morphine. Only when it is clear that you are too out of it to experience any struggle to breathe will the breathing machine be turned off. I have always said I am not afraid of dying -- I just don't want to be there when it happens! This seems like the way to accomplish that.

## *Equipment Cost*

Insurance policies vary in their coverage of medical equipment. Talking to a respiratory equipment supplier in your area. Your insurer may be familiar to them so they may be able to tell you what is generally covered but your insurance policy is the best source. It is important to be certain that the DME (Durable Medical Equipment ) provider is covered by your insurance if your insurance requires you to use "in network" doctors and other providers. If your copy of the policy isn't specific you may want to call the insurance company. If you call them, document what you are told, the date, time, names and job title of the person you talk to, and take notes.

By the time you need a ventilator you will be eligible for Medicare (if you have paid into Medicare). The tricky part is whether Medicare will be your primary insurance (pays first and any remaining amount is then filed with any other insurance you have) or if it is secondary (filed after your other insurer has paid their allowed amount.) If you are covered under a company policy of someone who is still working, Medicare will be secondary. Medicare will be primary if the policy holder is retired even if their retirement package includes a Medicare Supplement policy.

If the monthly out of pocket expense is obviously not affordable, ask the vent supplier if they have a financial assistance program. Apria has one and to apply for assistance you need to fill out a full report on income and expenses. You may be eligible for reduced rental or even no rental fees.

The best I can do is give you my cost. (Remember, this is my cost and every equipment supplier and insurance is different!)

I am on Medicare and have a Medicare supplement policy.

The vent rental and maintenance is billed at (Gasp!) \$3775 per month per ventilator.

*Ignore this ridiculous number.* Providers are free to bill Medicare any outrageous amount but Medicare will only approve a part of that and the provider has to accept with that amount. Suppliers who accept Medicare patients are not allowed to come back and bill you for any more than your 20% Medicare copay!

Medicare allows \$896 of which they pay 80% or \$703. That leaves me with a monthly bill of \$179.

My supplemental has an "out of pocket" \$3000 yearly minimum so unless I have other claims, they pay nothing. Two vents would cost \$358 per month for 9 months if I don't have other claims to reach the out of pocket sooner.

Vent supplies (suction catheters' replacement hoses, filters, etc.) are not included in the monthly rental. The same idiocy applies to these items. They bill Medicare over \$500 for 75 suction catheters and I end up paying \$36. And I can get them for less than that online!

It is possible to purchase your own vent but you will not receive any of the services provided along with vent rental. When you rent, a Respiratory Therapist visits frequently at first, then less often. The therapist does no patient care. The therapists are there to check the equipment, not The patient. But I have any problems, the therapist can intervene to work with the doctor to change settings or recommend other equipment. During visits, the therapist will run checks on the vents, suction machines, batteries, chargers or other equipment. They arrange for very prompt replacement if needed. They keep records of all the machines so they are regularly exchanged and sent in for a full "tune up". They are on call 24/7 for equipment emergencies. Equipment is replaced at no charge if there is a problem with it. We order our supplies by phone through Apria as well and they are very prompt with most items.

## *Trach Care: The Rest of the Story*

In the process of making the decision of whether to have a tracheostomy tube placed you may hear myths about how complicated trach care is, how time-consuming, expensive, ugly, and problem prone trachs are. Worse, you may be told you will never be able to speak or eat again. Your doctor will focus on what a trach tube will do to help you and what the surgery will be like. After surgery, your nurses will teach you and your care givers how to care for a trach. (Well, sort of. You get postoperative trach care.) This article is intended to tell the rest of the trach story by correcting misconceptions and filling in some of the blanks about actually living with a trach.

### *Trach Care*

In the hospital trach care is performed and taught with a ritualistic precision rivaling a Japanese Tea Ceremony! Eventually, some nurse will let the cat out of the bag and mention that home care procedures aren't as strict. So what is safe and reasonable for home care? Once the trach incision is healed and no blood or blood tinged drainage is seen, here are some guidelines for care:

#### *Cleaning Around the Trach*

This part of trach care is done in 30 seconds as part of bathing or washing up. Once a day cleaning is enough unless you have a lot of mucus drainage around the tube or recurring infections. Gloves are optional for washing the area but good hand washing first is not.

For decades 3% hydrogen peroxide was the standard solution used for wound care. Today we know that peroxide damages the fibroblast cells that provide the scaffolding for wound healing, actually slowing healing and leading to the growth of granulation tissue (discussed later). It has also been found to have minimal antibacterial effect. Peroxide is very good for removing dried blood, drainage, and mucus, but once the tracheal incision stops bleeding it is time to move on to a different wound cleaner.

Finding a safe, easy to use, and inexpensive cleaner for long term use is easy: Buy some ordinary hand soap. Soap has as much antibacterial action as peroxide and none of the cons. A soap labeled as antibiotic is not necessary, in fact, these soaps kill only some bacteria and may allow other bacteria to take over. Additionally, this overuse of antibacterials is of concern to medical people and environmentalists. Finding a soap without antibacterials, lotions, or perfumes is not critical for trach care and is increasingly difficult, but they are available. You may be able to find Ivory or Jergens without antibacterials or check natural health stores for other brands.

What about the sterile water, 2x2's, Q-tips, and ointments and gloves sent home with you? By the time the first batch of these supplies is used up, the edges of the wound should be well healed and plain soap and water can be used. The 2x2's and Q-tips can be replaced with clean wash cloths. Buy a big economy pack of cheap, thin wash cloths. Plush wash cloths are too thick to get in close around the trach.

The best thing for the skin around the trach is exposure to air by skipping any ointment and, if a 4x4 is used, keeping it dry. Most people continue to use the split 4x4s around the trach to absorb any drainage, reduce air leakage and to hide the skin opening (stoma). Unless an infection develops, an antibacterial ointment isn't needed. If the area seems damp all the time, an ointment containing zinc oxide provides a good moisture barrier. An itchy, burning trach site may have "athlete's foot" from being warm and damp. A couple days use of a non-prescription anti-fungal cream, combined with twice a day washing and keeping the area dry will clear it up quickly.

### *Inner Cannula?*

A trach tube with an inner cannula is actually a tube within a tube. The smaller inner cannula is removed for cleaning daily. Some trachs come with disposable inner cannulas which make trach care fast and easy but expensive. The need for an inner cannula varies and not all people with trachs have one. See *Trach Choices*, Page 70.

Because the inner cannula is inside the body and warm and moist, this isn't a good place to cut corners on cleaning. Continue with the sterile procedure as taught. Or ask your doctor if you can be switched to a trach without an inner cannula. That is a big time saver!

Trach cleaning kits are handy and include a brush that fits inside the inner cannula, but to save money the brushes and containers can be washed and reused. The trick here is thorough drying after washing any equipment to be reused. Putting them away damp provides an excellent opportunity for bacterial and fungal growth.

Daily changing of the trach tie isn't needed. The most economical trach tie is a roll of twill tape. More expensive types of trach ties have the great advantage of allowing easy adjustments to the tightness of the ties because they use a Velcro closure. These ties can be washed and reused many, many times before the Velcro loses its grip.

### *Suctioning*

The first question asked about suctioning is "Does it hurt?" No. The coughing it causes may look downright lethal to observers but doesn't hurt. In fact, suctioning almost feels good because it gets rid of the congestion so easily. A half minute back on the vent to catch your breath and all is well. A caregiver may ram the suction catheter (tubing) in too far and that does hurt, but doing it correctly (inserting the catheter just until a cough is triggered, not until you hit bottom) should not be painful or even particularly uncomfortable.

For the person suctioning, the difficulty is not in the actual suctioning. That is simply a quick vacuuming. The challenge is in getting a glove on, the catheter out of the package and connected to the tubing, the machine on, and the trach hose off -- all without allowing the catheter and gloved hand to touch anything but each other! It seems like learning to juggle at first but quickly becomes as automatic as tying shoe laces. Anyone -- friends, neighbors, kids -- can taught how to suction with a simple explanation as they watch it done and the opportunity to practice.

Suctioning rarely takes longer than 3 minutes for set up, suctioning and clean up, but sometimes several passes with the catheter and time to catch your breath in between are needed. How often suctioning is needed varies widely. Some patients are suctioned only two or three times a day, others a dozen or more. Every one has good and bad days too.

Caregivers, don't try the "I will hold my breath to determine how long my patient can be off the ventilator" trick. You will take a deep breath first. Your patient can't. He or she can't hold a breath-- it escapes out of the trach. On top of that, you are about to suck any remaining air out of their lungs! Tolerance for suctioning varies so just watch for any sign that your patient is feeling suffocated and put them back on the vent for at least a few breaths before suctioning again.

Just as with trach care, home suctioning is simplified. Someone prone to respiratory infections will want to be more strict. The full sterile method used in the hospital requires a bottle of sterile water and a sterile suction kit containing a suction catheter, a pair of gloves and a small sterile container for the sterile water. Nurses may want to brace themselves as I describe the way we do suctioning. It is NOT the way you were taught! Safe? It has worked for me for 12 infection free years! Suctioning can be done with just one non-sterile disposable glove (not reused). At home, this

can be simplified unless you have a parade of different caregivers. My routine has been pretty much whittled down to "Don't touch the business end of the catheter with your bare hands." And "There is no five-second rule if you drop it!" The steps are:

1. Loosen the trach hose so you won't be fumbling one handed to pry it off.
2. Open the package or baggie containing the suction catheter.
3. Put on one exam glove (not sterile but from the manufacturer's box). Pick the glove up by the cuff with one hand and pull it on the other hand using the cuff to tug it up. The idea is to get it on without touching anything but the cuff with your other hand. You now have one clean hand and one which you will now consider absolutely filthy with germs. (It is!)
4. With the gloved hand, take the suction catheter out by the suction tip, not the connection end, winding it around your gloved hand.
5. With your ungloved hand, attach the suction catheter to the suction machine hose and turn on the suction machine.
6. With the ungloved hand, silence the vent alarm and disconnect the trach hose from the trach tube.
7. Suction by covering the "chimney valve" with your thumb and inserting the suction tube into the trach. Insert it just far enough to trigger a cough. It really helps to rotate the suction tube with your gloved hand as you suction and gradually withdraw the suction tube.
8. Reattach the vent to the trach.
9. If the person needs more suctioning, wait until the vent gives several breaths, disconnect the vent and suction again.

A new catheter is used daily. Insurance usually limits the number of catheters used per day. Medicare allows 3. In between uses, the catheter can be put back in the package. This is much easier with "straight packaged" catheters than coiled ones. We find that zip lock baggies (changed daily) work better for coiled ones. The suction catheter itself doesn't need rinsing between uses, but if you want to, suction sterile water from a sterile container through it. Suction kits with pop up sterile containers are available for this. We put the baggie in the refrigerator with the idea that the cool temperature will slow any bacterial growth. I don't know if that actually works, but without a designated place to put it, we would probably go through our allotted 3 per day every day just because we couldn't find the open one!

The suction machine tubing can be rinsed through by suctioning up fresh tap water (through the suction machine hose, not the suction catheter) from a clean glass. My husband and I don't like seeing a suction canister with "stuff" in it, so instead, he removes the suction machine hose and runs water through it after one or two suctionings. (Under the bathroom faucet, cold water, and stretched to drain into the toilet, not down the sink drain. Yuck!) This way the "stuff" never reaches the canister. We don't bother with this at night though. The canister gets rinsed out in the morning.

### *Trach Changes*

The first trach change is generally done before you leave the hospital. After that it may be done in the hospital's out patient clinic, doctors office, or by Home Health. When you call for a trach change appointment tell them the brand, type, and size trach you have (i.e. Shiley, cuffed, size 7) so it can be ordered if necessary. We order my trachs through our vent supplier, and they are delivered to our home. That way we always have the right trach on hand for a home emergency. (In 13 years we have never had to replace my trach at home.) It is a very simple procedure that anyone can do



and unless there are problems with an abnormally shaped trachea or granulation tissue (discussed below) most trach changes can be done at home by caregivers who have seen it done several times. Caregivers should at least assist with a trach change to be prepared for an emergency.

Coughing is perfectly normal for several minutes after a trach change. The trachea is made to react with strong coughing any time it is messed with. I always thank the RT changing my trach before the change because I know I will be coughing too much to say it after!

If you have bleeding with trach changes, it can be due to granulation tissue. If the bleeding is from the skin opening, try a bit of hydrocortisone cream around the trach once daily for about three days before the trach change. That will shrink the granulation tissue and reduce the bleeding. If the trach change causes deeper bleeding that results in blood in the mucus suctioned after changing the trach, that may indicate granulation tissue down in the trachea. This necessitates more frequent trach changes, not less, to prevent the tissue from building up and making changes more difficult. A consultation with an ENT doctor should be done.

Frequency of changes varies from weekly to monthly, to every other month, to "whenever it seems to need it." People prone to granulation tissue will have easier changes if they are done frequently and frequent changes may reduce respiratory infections, especially in people with sinus problems.

### *Trach Choices*

Once your trach tube is put in you may think that it can't be made better. You don't have to continue with an off the rack, standard issue tube, however. Options include:

- ◆ Specially sized trachs for people with very long or short necks are available as well as custom fitted trachs for anyone whose trachea is abnormally curved.
- ◆ With or without an inner cannula.

The purpose of the inner cannula is to allow easy cleaning of any mucus that sticks to it and builds up. It also provides a fast way to remove complete blockage by simply removing the blocked inner cannula and sliding in a new one.

Newer trach tubes, such as Bivona trachs, have a silicone lining that prevents mucus from sticking to the tube and building up eliminating the need for the inner cannula. However, someone with frequent large mucus plugs that are not easily removed by suctioning would need the inner cannula so that it could be replaced quickly if plugged up -- if the plug is actually in the cannula and not just below it or down in the bronchi.

Using a trach with an inner cannula after a tracheostomy allows time to evaluate whether the patient needs the inner cannula. It can be replaced with a trach without an inner cannula with any routine trach change. A trach tube without an inner cannula is a simple, no fuss trach that eliminates cannula cleaning.

- ◆ A TTS (tight to shaft) trach is an option that is especially helpful for those who have difficult trach changes. A TTS cuff flattens tightly to the shaft of the trach tube when it is deflated, making tube changes easier and more comfortable.
- ◆ Cuffed or uncuffed trach.

A cuff allows better control of the volumes of air given by the vent. With the cuff inflated, the full volume of air is delivered to the lungs. Deflated, a significant portion of the air escapes through the nose, mouth, or around the trach tube at the stoma. Patients who don't have a lung disease such as emphysema or COPD generally have no problem with a deflated cuff or cuffless trach.

An uncuffed requires a stable respiratory status. I find I need a cuffed trach. Even though I have the cuff deflated during the day so I can talk, I need it inflated for sleep, or I snore horrendously!

- ◆ Another trach option is a trach which has two suction ports, one to suction the lungs and another much smaller one that allows suctioning of the area just above the cuff. This allows above the cuff suctioning right before deflating the cuff to remove fluids waiting to drop into the lungs. The downside is that this second suction tubing is part of the trach itself and is so narrow that it is easily clogged making unusable until the next trach change.
- ◆ Fenestrated trachs have a hole or holes along the tube to allow some air to escape and travel the normal route up and through the vocal cords. This allows the person to speak. Fenestrated trachs can encourage the growth of granulation tissue around the holes and cause bleeding with trach changes. Other methods of providing air for talking are recommended.

### *Speaking with a Trach*

- ◆ For people who were able to talk before being trached there are some options for resuming speech. The first thing to try is simply deflating the cuff so that some air can pass over the vocal cords. If you can talk with the cuff open, you will find that adding about 5 of PEEP (Positive End Expiratory Pressure) to your ventilator by adding a PEEP valve to the tubing will give you the reserve air volume to speak more smoothly rather than saying one or two words with each breath. For some reason, adding to the PEEP settings on the vent itself doesn't seem to work. An external PEEP valve inline on the hose does. The volume delivered per breath will probably have to be increased to make up for air lost around the deflated cuff. For example, my volume is set at 700 and with the cuff deflated about half that goes up through my vocal cords rather than into my lungs. The remaining 350 cc's is plenty for comfortable breathing.
- ◆ Another option is a speech valve such as a Passey Muir that directs exhaled air through the vocal cords. These valves also require that the cuff be deflated during use so may need volume setting changes. A speech therapist will show you how to use it.
- ◆ There are also "talking trachs" that use compressed air, rather than air from the lungs, to pass over the vocal cords. Because they require an external source of air they are less convenient, but for anyone whose breathing can't tolerate any air loss from the lungs, these are an option.

### *Eating and Drinking*

As with talking, if you could swallow safely before the trach you should be able to after. Although a cuffed trach can provide some protection from food and liquids getting into your lungs, it isn't complete protection. Anything that "goes down wrong" when swallowing will simply sit on top of the inflated cuff and wait until the cuff is deflated to continue its journey to the lungs. Closing the cuff when eating can actually worsen the ability to swallow safely because it reduces the movement of the valve that closes over the airway during swallowing.

Your sense of taste and smell may be diminished somewhat when you have a trach because the air isn't passing through the nose, but not necessarily lost. With my trach cuff deflated, my sense of taste is, unfortunately for my waistline, unimpaired. My sense of smell is limited only in that I cannot sniff well enough to smell something being waved under my nose. A scent that permeates

the air such as food cooking, a vase of flowers, or something going bad in the fridge is very obvious to me! With the cuff inflated, my sense of smell is quite diminished but not erased, and my sense of taste is just fine.

### *Bathing*

One of the most often asked questions is "Can I take a tub bath or shower with a trach?" The answer depends on the individual. The last thing anyone needs is to get soapy water in their lungs! If bathroom space allows, you can position the vent alongside the tub or shower and protect the vent from splashes with a plastic drape or bath towel. If you can tolerate having the trach cuff deflated and the trach plugged while you wash, there should be no problem.

If you can be off the ventilator but can't plug the trach, a shower is out and bathing requires caution. Bathing using a tub or shower sling prevents the risk of slipping under the water but the trach still needs to be protected from sprayed or splashed water. Soaking in a tub or shower feels good but certainly isn't necessary for keeping clean. Being moved to a shower or tub chair, then back again is a lot of work for your care giver and tiring, uncomfortable, and chilly for you. A "sponge bath" is faster and easier on you both. But don't use a sponge. Use a wash cloth and scrub, don't just wash. A delicate touch won't remove flaky old skin, but some areas may be too sensitive for scrubbing!

### *Unexpected Aggravations*

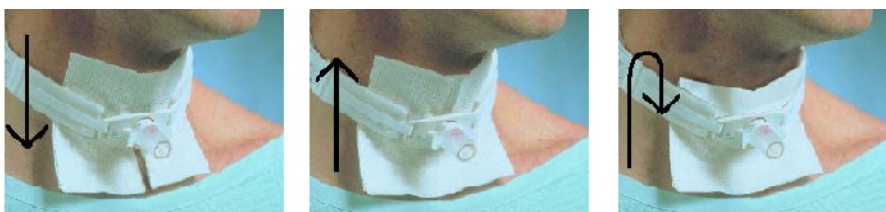
Many of the day to day aggravations of life with a trach are never mentioned by medical personnel. They don't tell you that you may have a rafter rattling snore if you fall asleep with the cuff deflated. They don't mention that your ability to smell won't work quite as well when you no longer breathe through your nose. Worse, you can't sniffle or blow when you have a cold! Other aggravations include:

#### ◆ Air Leak

Perhaps the most annoying non-medical problem of a trach is air leaking around the trach and out the stoma. If you are on a vent the air loss can be enough to set off the alarms but generally, it just causes noisy whistles, gurgles, or whooshes of air with each breath. Of course inflating the cuff will solve the problem but it will also prevent talking. The best solution for an air leak that is driving you nuts may be to increase the size of your trach with your next trach change, but here are some things to try first:

- ✓ Sometimes just re-adjusting the trach so it is centered helps.
- ✓ Although tightening the trach ties seems like it should help, loosening them is actually the thing to do. Loosening the ties lets the trach tube sit at a slight downward angle which blocks the airway a bit more. Tightening the ties may help if they were loose to start with, but too tight ties, especially narrow ones, will lead to pressure sores under the ties.
- ✓ Trach leaks occur when the vent hose is allowed to hang to one side, pulling the trach a bit off center. There are lots of ways to rig up a little support for the hose. Duct tape, rubber bands, PVC pipe, and safety pins have all been put to use by trached vent users! I spend most of my day at my computer, so I have a spring arm on one side to loop the hose over. A flexible arm desk lamp with the lamp and wiring removed is an inexpensive version of a spring tension arm.
- ✓ After a lot of trial and error (more gauze pads, fewer gauze pads, cutting the pad to open the slit to a trach sized hole in the middle, and stuffing Kleenex into leaky spots) I finally

found a method that has greatly reduced my air leaks. Put one split gauze pad (drain sponge) around the trach with the split pointing down. Place the second one on top of that with the slit pointing up. Then fold the top edges over and tuck the outer corners under the collar and trach plate. This also gives a neater appearance -- no gauze flapping around or tickling your chin.



### *Discomfort from hoses*

The stiff plastic of vent hoses makes it hard to position the tubing so that it is not pushing or pulling at the trach. To reduce this you can get a short length of a more flexible rubber hose to put between the trach and the vent hose. This rubbery hose is used in the set up for nebulizer treatments so should be available from your respiratory supplier. This softer hose is more comfortable, but acts as an echo chamber for any rattly congestion!

### *Coughing*

Another aggravation is that whenever the trach or vent tubing is moved it sets off a coughing spell. You quickly get past the fear that it causes and recognize that it is just an aggravation and not a problem. There isn't anything to prevent this, but simply disconnecting from the hose when transferring or being turned in bed really helps. Most people tolerate a minute off the vent with no trouble.

### *Granulation Tissue*

Granulation tissue is an overgrowth of tissue as the body attempts to close a wound. It can grow out around the trach opening, looking lumpy, bumpy and red, purple or pink because it is full of blood vessels. It leaks serosanguineous fluid making the area moist and a haven for bacteria. It can bleed easily, can become infected, and may be sensitive and uncomfortable. It can get in the way and make trach changes difficult and may even develop down in the trachea causing bigger problems.

Not everyone seems to be susceptible to developing granulation tissue and it is unclear what triggers it. There is some evidence that continued use of hydrogen peroxide for cleaning causes it, and the logical suspicion that frequent trauma to the trach site and the trachea itself from pulling on the trach and tubing will stimulate it. Since the cause is unclear, suggestions for prevention are limited to not using peroxide and stabilizing the trach tube as much as possible.

If granulation tissue appears at the trach site, applying cortisone cream is usually effective in shrinking it, or silver nitrate can be used by your doctor to chemically burn it away. Another possible home remedy is a cream containing zinc oxide. Keeping the area clean and dry is especially important if the granulation is leaking fluid. Granulation tissue down in the trachea can usually be removed with laser surgery.

## *Caregivers for Vented PALS*

When someone is considering going on a ventilator one of the first concerns is the added work for the caregivers. The amount of time added varies with the frequency of suctioning and that generally decreases as patient and caregiver adjust to the vent, get organized, and learn that much of what they were told needed to be done is for hospital care, not home care! In addition to suctioning there will be daily washing of the trach area and, depending on the type of trach you have, cleaning the trach tube. Cleaning suction equipment is daily. Changing vent hoses and filters generally drops from weekly to whenever unless infections are frequent. Ordering supplies should only need to be done once a month at most. Any significant increase in care needs is far more likely to be the result of advancement of ALS weakness rather than going on a vent. After a period of adjustment I think you will find your caregiver is actually less stressed, at least by the breathing aspect of your ALS. Congestion that now leaves you both wondering if you can cough it out or if you are going to die this time will be nipped in the bud with suctioning. Any choking spell can still be exhausting but the trach and vent will assure that your airway can't be blocked and you will continue to breath throughout the choking spell.

The reality is that the extra daily workload is manageable but the added months or years of care may not be.

Many people are told that they will have to hire round the clock nurses. Big lie! Or they are told that they will have to hire a vent qualified nurse anytime your caregiver leaves the house. Also not true. If all you need is someone to give your main caregiver time out of the house for shopping, errands, appointments, or just plain escape. Not a nurse, just a relative or friend or two willing to learn how to suction you--and that is nothing more than a minute of glorified vacuuming--and the basics of the vent. You have to ask. No one is going to know that you need help unless you tell them!

The company supplying your vent may offer a training class for anyone willing to help but that isn't a requirement. In fact, they may scare off volunteers with a lot of technical jargon! You can show someone the ropes in ten minutes of instruction and demonstration plus time for them to practice suctioning you. The learning curve is not steep! Vent basics include deciphering vent beeps. If the vent beeps you either need suctioning, a hose is off or loose, or the battery is low and the vent needs to be plugged into a wall outlet. If the problem isn't found they just need to use the ambu bag while waiting for the vent supply company to show up. Suctioning is simple vacuuming. The hardest part is getting a glove on and then remembering not to touch anything except the suction tube with that hand! Most reasonably coordinated people can be shown the procedure, practice it a couple of times, and be ready to go.

If you want a home health care agency to provide care, that can be slow to get started. Check with available agencies early in your planning. Most agencies don't have many, if any, nurses trained and legally allowed to care for vent patients. Even if they are, their visits will probably be restricted to an hour or two once or twice a week. You can get aides to help with bathing and getting you out of bed etc. but someone else must be there to assume responsibility for the vent care. They won't be allowed to give tube feedings or medications either. LPN's and RN's can do those things even if they aren't allowed to be responsible for vent related things. Because of the legal liability limits of Home Health Agencies, the cost, the rather frequent problem of them not showing up, and the possibility of getting someone you just don't want caring for you, friends are most often a better solution for shorter periods of care. If their visits are scheduled on a regular basis it is much easier for your regular caregiver and your other helpers to plan ahead.

The handbook, "Share the Care" is a guide to finding and organizing a group of helpers to form a strong working support group for people and families overwhelmed by care needs. The "Share The Care" handbook is available for you to purchase from Amazon or at Barnes & Noble online and at bookstores for \$16.

Lotsa Helping Hands gives a place where volunteers and paid help can see your calendar, view requests for assistance such as time for shopping, cleaning, cooking, appointments, view gaps in care coverage, and sign up to help when needed. They can sign up for a single spot or schedule themselves for regular help.

Another option is to hire your own caregivers. As long as they are not RN's, LPN's, or CNA's, they won't be risking losing their license if you train them to do vent, meds, and feeding tube care. The problem here is the same as finding a sitter for little kids; safe, motivated, compatible personality, adaptable, honest, no criminal background, reliable. If you put out an ad for someone, don't include details beyond "home care help needed". Do background checks, ask for five or more references for work history and personal attributes -- and follow through on them. A good source of caregivers are local nursing or other health care schools (they don't have a license to risk until they graduate) or volunteers from your church.

A live-in caregiver can be great even if they are also working at another job. You can negotiate any pay expected so this saves money. A live-in is a much more personal arrangement so it definitely requires the right personality as well as passing the other checks. It also requires laying out of non-care rules such as smoking, pets, visitors, groceries, etc.

Some states will now pay a small hourly wage to family members who provide your care. They realize that whatever they pay is far more cost effective for the state than forcing you onto Medicaid and into a nursing home.

So, Home Health is not required as long as you have enough family, friends or privately arranged caregivers trained to manage basic vent problems and suctioning. Or, you can use Home Health at first and once you are settled in at home you can train your own people (they definitely don't have to be nurses!) and let Home Health go.

### ***Planning for a Vent***

If you have decided to go on a vent when the time comes, don't let the time sneak up on you. A planned and scheduled switch to trach and vent is stressful but an unplanned switch is worse. Even if you aren't ready to set a date, the information here can help you be one step ahead, especially if the vent event takes place sooner than anticipated. Surgical planning and home preparation can be done without scheduling anything. It is actually quite a bit like having a baby. You know that it will happen but not precisely when, and you don't wait until the baby arrives to get a crib and diapers! So here are some practical tips to help you be prepared for the day you bring home your bundle of joy, little LTV 950 or precious Trilogy.

When you meet with your Pulmonary doctor to discuss going on a ventilator, request that an ENT (Ear, Nose, and Throat) Surgeon put the trach in. It isn't as easy as Father Mulcahey, Radar O'Reilly and a pocket knife made it look. A bad trach job will be a problem that may end up requiring corrective surgery. An ER doc won't do a trach unless your face is smashed. He will put an endotracheal (mouth to trachea) tube in, put you on a vent, admit you, and let your regular doctor arrange for a surgeon/ENT to schedule the trach. If you have pneumonia, they will do the same and wait until that is under control before doing the trach. An endotracheal tube is wretchedly uncomfortable -- another reason to have a trach put in before you end up in the ER!

Assuming you are following my “Do as I say, not as I did” advice, you will also ask to meet with the ENT surgeon or his nurse to discuss and see the type of trach he will put in. Seeing an actual trach will go a long way in clearing up any confusion and be of great help later in caring for your trach. Knowing what is hidden from view makes it all easier to work with. Don't worry at this point about the brand and type of trach that will be put in. That can be changed in a few weeks during a trach change.

If you don't already have a feeding tube, this is the time to have one put in even if you don't need it just yet. Aside from being handy for a fast, minimal effort meal at times when your caregiver needs a little break, avoiding another hospital stay or even an outpatient procedure is a real plus.

Ask your doctor how long you will be in the hospital and if you will have to be sent from there to some type of facility where vent and trach care training is provided or if that is done at the hospital. This is also the time to ask about the availability of nursing homes that take ventilator patients. In spite of your planning to remain at home, if your primary caregiver can no longer do the job, a nursing home may be your only option short of discontinuing the vent. Nursing homes licensed to take vent patients are currently few and far between. Your only option may be quite far from your family.

Find out from your doctor who will be supplying your ventilator and other equipment. If your doctor doesn't know which type of vent they supply, call them and ask. Then call the company you got your wheelchair from and explain that you will need a vent tray or mount added to your chair. They will need to know the type of vent in order to get the right mount. Hash out the details of filing the Medicare/insurance claim. They may not be able to file until you have the vent which is fine but a nuisance using your chair with out the vent mount. With bulbar onset ALS, a vent may be needed while you are still able to walk. A vent is too awkward and heavy for anyone with any upper body weakness to carry in a backpack but a folding grocery cart may work.

Expect to be in Intensive Care after surgery. You aren't going to be in such bad shape that you need Intensive Care but few hospitals have any other area that has nurses trained and allowed to care for ventilator patients. That same requirement will land you in Intensive Care for anything in the future, even a simple appendectomy.

Line up volunteers to sit with you at the hospital if allowed, and especially if you are moved to a vent training unit. The nurses are never staffed well enough to make a newly trached patient feel safe and having someone with you is reassuring even if all they can do is run out and grab a nurse.

Ask for a soft touch hand or pillow call button. Getting everyone to remember to make certain you can press it before the leave the room is a big problem though. Feel free to raise hell up the line if it isn't done. A vent alarm response will be frightening slow for panic prone newby -- and we are all panic prone newbies at first.

There is a suture on each side of the trach at first and they can get tugged on by the vent hose when you move. Moving also tugs on, moves, or jiggles the trach. That triggers the cough and gag reflexes and, although not painful or dangerous, it is scary at first and forever annoying! You can skip all that drama by taking the vent hose off the trach when you are repositioned in bed or transferred to a chair. You will be pleasantly surprised at how long you can do without the vent now that you are properly oxygenated to start with!

If you don't want to be held prisoner for a month, you may have to push. I spent over three weeks in the special care facility that was supposed to get me ready to go home, and even though I had absolutely no medical complications, NOTHING was done until the last week! I suspect that this was all due to the fact that Medicare would automatically cover 21 days and the facility wanted

every dime of that even though I could have been home in a matter of a few days. Anyway, make certain there is a planned time frame for getting you home -- and that it is in writing and required reading for all RN's. Get them moving right away on obtaining your vent rather than the hospital supplied one. Schedule vent training classes for your care givers ASAP. Ditto for suctioning and trach care instructions and hands on experience for your caregivers.

Include your preferences in your discharge plan. If you don't let them know, you could be sent home with a urinary catheter still in, and on tube feedings even if you can swallow safely. You will probably have a swallow study before they will let you eat.

If you were able to talk before the trach, you will be able to talk after. It won't help with problems speaking clearly but you will be able to speak more loudly and without exhaustion. A Speech Therapist will probably want you to try a Passey-Muir Valve. Not necessary. Ask the doctor to deflate your trach cuff, wait until you stop coughing, then try to talk. If you get short of breath with the cuff deflated, the solution is to have the vent setting changed to a higher volume to make up for the air you lose with the cuff deflated. Then call someone and make them cry at hearing your voice again.

If you have been getting out of bed at home, get out of bed as soon as your doctor allows it. Don't stay up to the point of exhaustion, but getting your strength back should be easy now that you are breathing properly. You will probably be far more comfortable if you have your wheelchair brought in.

If anyone seems to think you should try being off the vent for increasing periods of time, feel free to tell them you have no intention of EVER being short of breath and miserable again. You will come off the vent just for the sake of knowing how long you are comfortable (could be a half hour or several hours), but there is no point in trying to extend your time off much less get you off the vent as they normally do with ventilator patients. Remind them that you have ALS so your breathing will never improve no matter how hard you try.

The combination of anesthesia and pain meds will almost inevitably cause constipation if a stool softener (Dulcolax, Colace, or Docusate) isn't taken daily, ideally starting a day or two before the trach is put in. It is critical that you do not begin eating or being fed through the tube until your intestines "wake up" from any anesthesia you are given during the procedure or surgery. For some reason they are slower to shake of the effects of anesthesia than the rest of the body. This applies to any procedure or surgery! There is some evidence that gastric motility, the passage of food through the stomach and intestines, is slower in ALS patients, but whether this is a direct effect of ALS on digestive muscles is far less likely than the effects of not being able to get up and walk! If you ever had surgery before ALS, you know that you will be ordered up and walking far sooner than you want to. The assumption is that it is to help you get your strength back. Not really. It is to get you breathing more deeply and get your bowels working.

If you are in the hospital during this time, your nurses should be listening to your lower abdomen with a stethoscope to hear the gurgling that signals the return of bowel function. Until then, you must not be fed. Severe problems with the intestines can occur in anyone after surgery and because of immobility, ALS patients are somewhat more susceptible.



## *Home Preparation*

### *Power Supply*

Having an adequate electrical supply is important when a ventilator will be in use. The DME (Durable Medical Equipment Provider) you get your ventilator from may send someone to look at your wiring before entrusting their expensive equipment to you. It is important to make certain that the electrical circuits in rooms where you will be using the vent won't be so overloaded that you will be blowing fuses all the time. A vent doesn't draw much, but a bedroom with a vent will probably include a lot of other equipment; When you start adding up all the electrical equipment that is likely to be used in the bedroom, it is surprising: lights, electric hospital bed, suction machine, nebulizer, electric blanket, fan, TV, CD Player, call system, electric or battery powered lift, and charging outlets for the vent, wheelchair, and every other battery operated thing that requires recharging. (Label every battery charger and cable with the name of the piece of equipment it goes with or you will end up in Charger Hell.) Thankfully not everything will be in use at once, but check what other rooms might also be on that circuit. Living rooms can be electricity hogs too. TVs, sound systems, computers and all their peripherals, lighting, space heaters and fans. Kitchens definitely would not be good companions on a circuit with either your bedroom or living area! Few houses have enough outlets in the right places for what you need to plug in so it is very helpful to add outlets or power strips.

A generator is good to have and essential if power outages are frequent in your area. It seems that every part of the world has its own power problems. If it isn't a blizzard, it is a hurricane. Throw in a flood, tornado, or power grid failure and we are all likely to lose power at times. Getting your name on a call list for the power company is minimally helpful. They can't restore power to your house until they restore your neighborhood. And they can't restore your neighborhood until they restore the portion of the entire power grid that is out. So buy a generator. A full house automatic generator is terrific, but a small inexpensive generator and a backup battery are all that is needed to keep a Bi-level Ventilator or ventilator running. Run the machine on one battery while recharging the other. The worst thing for a generator is not being used. The carburetor will gum up from the fuel sitting in it and it won't start. In addition to the seasonal checks, run it briefly every month or so.

### *Storage*

You will have lots of supplies on hand and will need a place to store them. A closet will need shelving, or buy a tall storage cabinet. You will probably be sent home with all the essentials (which, in time, you will discover includes junk you won't need once you are out of the germmy hospital environment). The essentials are the vent, suction machine, and suctioning supplies, an ambu bag, back-up trach, and rechargers and cables for each machine. You will probably be sent home with a nebulizer and meds for breathing treatments. They generally aren't necessary but are ordered anyway. Good to have on hand for times of increased congestion though. Ideally there will be a backup vent. Medicare coverage for a second vent generally requires that you use a different type of equipment at night. More critical than the backup vent, however, is a backup suction machine. If the vent should fail (extremely rare) you can use the ambu bag, but if the suction machine fails when you are getting plugged up, there is no time to wait for a replacement to be delivered.

A bedside table or cart will be needed to keep suctioning supplies handy. A cart is nice because it can be moved with you to the living room. Make an effort to avoid turning your home decor into hospital tacky. Equipment can be in closed cabinets. Visitors don't want to see all that stuff and you want to keep your home as normal looking as possible. If you live in a hospital environment you will feel like a patient and you are not a patient once you leave the hospital!

### *Beep Beep Beep!*

A ventilator will alarm if the volume of air being delivered drops below a set level, such as when the hose is disconnected from the trach or vent, or excessive air is leaking around the trach. It will alarm when the patient is getting congested and not enough air can be pushed in. If the battery is getting low, it will alarm but continue to run until the vent's internal battery is depleted. In practical terms that means beeping whenever you are suctioned, popped off the vent for a turn or transfer, or the power source is changed. The alarm can be temporarily silenced by pressing a button, but over time you begin to just let it beep for the couple of minutes it takes to suction or whatever. That “beep creep” leaves your caregivers somewhat deaf to the sound!

The huge problem is that although that alarm sounds so loud to us, it isn't always loud enough to alert a caregiver who is sleeping in another room, watching TV, showering, doing laundry, vacuuming, or outside. Bottom line is that you need a second way to get their attention and that second way must have good range and a beep or vibrate option. There is a big lack of this technology available. In the past, it was fairly easy to adapt a wireless door bell for use with a capability switch that can be activated with only a small amount of pressure. Unfortunately, nearly all the wireless door bells today have lithium batteries which cannot be adapted. I have put together a list of call systems that can be used with a capability switch. It is a short list! Go to [Call Systems for ALS Patients](#)

### *Oximeter*

It is helpful to have an O2 Saturation fingertip monitor while getting used to suctioning. It can help you see what you can tolerate and learn what is normal for you. That is important because it is really helpful in figuring out if you are in respiratory trouble if you don't feel right, seem drowsy or confused, or have a cold or increased secretions. An oximeter doesn't have to be fancy or expensive. Renting from the vent supplier is over priced, adds to your copay, and over time adds up. Just buy one. Before you buy one from a medical supply company, check online at pilots supply companies. They may be considerably cheaper and will do the job just as well.

### *Home Again*

It takes a while to get over the heebie jeebies of the whole vent thing, but once you are settled in a bit, there are a couple of things you can do to simplify care. This process will take a little time and will happen as you become comfortable with care routines and as your trach and feeding tube stomas heal. (Well, they never actually heal because your body will always want them closed!) The same relaxing of rules applies to your vent. There are no official rules for how often you change the hoses (circuit) and filters when you are at home. Hospitals may require daily or every other day changes. That is not needed for home care and would be horrendously expensive. Try a two-week change and, unless you are very prone to pneumonia or have a big problem with mold growing in the hoses, you can stretch it out to monthly. If you use a humidifier on your vent, stick to frequent cleaning of that equipment as mold and fungus love humidifiers.

First, begin to separate the stuff you were taught about trach and feeding tube care from what really needs to be done! What you were taught is hospital protocol and although it is necessary for a germy hospital and post op care, it is not necessary or realistic for home care unless you have a constant parade of different caregivers. Refer to Trach Care: The Rest of the Story, Page 68, for more info on hospital care versus home care.

Make a list of supplies that will need to be reordered. Include the manufacturer's name for it, product number, how many per box or case, who to call to reorder, and phone number, space to enter date ordered and number ordered, space for date received. This will help you keep track of how much stuff you use so after a couple of months you will be able to order a two or three month supply all at once. You will see which items take more time to be delivered, and hopefully won't end up with a two year supply of anything to find space to store because you ordered 10 cases instead of 10 boxes!

Put together a medication record and add lines for vent maintenance. Add a copy to the packet you take along to medical appointments so you have a list of your meds and dosages with you.

### *Out and About*

Now that you are home, it is time to leave! With the vent, you should have more energy and less anxiety so going places will be easier. Your trach will draw far fewer stares than your Bi-level Ventilator headgear did -- and you will be pink and healthy looking! For travel, you will need a travel bag for the suction machine. Mine came in one and has room for suction kits and other stuff such as Kleenex, straws, etc. That is all you really need for short jaunts, but I strongly recommend keeping a adapter cable in your vehicle that can power the vent from the cigarette lighter/power outlet. Also, keep a second ambu bag in the vehicle. Don't ever count on remembering to bring the one you have in the house along! You won't! Whenever we are going to be gone for a few hours, my husband insists on bringing everything but the kitchen sink. We take my back up vent in a carry on size suitcase and always keep a towel for spills, sweater for air conditioned rooms, and a second urinal in the van so there is no forgetting the essentials.

## ***Safe Harbor: Rediscovering Life; Going on a Vent***

*by Diane Huberty*

2005, MDA ALS Newsletter

This month marks my first anniversary of life on a vent, life with a tracheostomy tube and mechanical ventilator. As I look back over this year and try to sum up my feelings, the words that come to my mind are "safe harbor". I have found a shelter, a place where I can literally and figuratively catch my breath, get my bearings, and even make some repairs to my storm-battered body.

The effects of the vent on my health, including oxygen levels that prevent headaches and give me energy, stamina, appetite, and pink cheeks, are equaled by improvements in my quality of life. My days are no longer dominated by shortness of breath, tiredness, and fear of the next episode of lung congestion.

My world has re-expanded along with my lungs. BiPAP (bi-level positive airway pressure, a non-invasive form of ventilation) was fine for relieving nighttime breathing problems, but as my daytime use of it increased, my world shrank. Even if the BiPAP machine had fit on my wheelchair,

the mask and headgear were too bizarre-looking for me to want to go out. As a result I seldom went outside, much less left home.

Simply talking left me breathless, dizzy and worn out, making socializing too much work. I needed 10 or 12 hours of sleep a night and still was exhausted by bedtime. Now, with the vent, I am out for hours shopping, sitting outside chatting with neighbors, supervising outdoor projects, even accompanying my husband to his part-time job and staying up too late! The vent doesn't draw stares like the BiPAP.

People talk about the horror of being "tied to a machine" when they talk about vents, but they've got it all wrong. I already was tied to another machine, my wheelchair, and like the wheelchair, the vent has only increased my freedom, not tied me down. I actually feel rather proud to be seen out and about doing normal things. I feel I am representing the physically disabled and showing able people that, vent and all, we are still more like them than different.

### *Feeling Secure*

Of all of the improvements the vent has made, the greatest is the feeling of safety it has given me. It sounds strange to say I feel safer with my life dependent on a hunk of -- egad! -- computer-run hardware and some batteries, but I do. It is far more dependable than my own body was and, unlike my body, comes with a backup unit!

The vent has alarms, something not all BiPAP machines have, which contribute to safety. There are no more horrible episodes of not being able to reach my call button when I can't breathe because of congestion or disconnected tubing. The vent alerts my family for me.

Life-threatening congestion is also a thing of the past. Relief is just a suction tube away! That little rattle in my chest that once heralded the onset of an exhausting and frightening hour of trying to cough out congestion is now just a signal that I need suctioning. Suctioning seemed like a big deal at first but quickly became fast and easy for my caregivers, a mere annoyance, not a medical event! Suctioning makes me cough hard so it probably looks like a miserable experience, but it is not at all. It is no more uncomfortable than any ordinary coughing spell and it leaves me rattle free. It is an incredible improvement over swigging expectorant cough medicine and then trying to hack up congestion using a cough-assist device or a series of Heimlich-like belly bruisers!

How does it feel to be on a vent? It doesn't! I am as unaware of my breathing as anyone else. No feeling of air being forced on me, no arguments with the vent over the rate and depth of my breathing. I just breathe! I expected a general ache or sore-throat feeling from the presence of the trach tube, but I am aware of it only if it gets tugged sideways or needs repositioning. At times the skin around the opening may get irritated, causing the only discomfort.

### *The Costs of a Vent*

What does it cost to be on a vent? Thanks to Medicare Part B, the cost is manageable. My vent costs include vent, nebulizer (to deliver medicine if I get a cold) and suction machine rental, disposable supplies (hoses, filters, suction catheters and glove trach dressings) and monthly trach changes. The cost of those supplies has gone down as we became more comfortable with less frequent hose, filter and trach changes, and switched to plain old soap and water for trach care. As far as we can figure from the slow Medicare/United Health Care/Apria process, we are charged \$283 a month for a single vent. Because we qualified for financial aid from Apria for 2009, we now pay only for supplies, about \$100 a month.

*2016 Update. Medicare now pays all except \$179 per month for my ventilator. They pay 80% of the cost of replacement hoses, filters, etc. I have found that for suction catheters, gloves and gauze squares. I can buy them online for less than my Medicare copay!*

(Added notes: This amount varies considerably depending on whether you have other insurance, who your equipment provider is, etc. A quick poll of a few other ALS patients on vents showed that most paid far less if anything at all! And, contrary to what many people are told by health care providers, no round the clock, paid professional nursing care is required! Family and friends can be trained at no charge by your respiratory equipment provider, and can provide all the care you need because of the ventilator.)

### *Burden of Care?*

What about the burden my care puts on my husband and friends? The only daily additions to my care are trach dressing changes and suctioning. Trach care takes about three minutes once a day. (I have a Bivona brand trach with no inner cannula [a tube within the trach tube] to change.) Suctioning takes about three minutes also, and the number of times a day I need suctioning varies from a few to a dozen. There are about two hours a week needed for equipment maintenance and an occasional nebulizer treatment to be done. All that doesn't add up to a lot of extra time, but when added to the hours of care I already require it does count. On the positive side, it is easier, faster, and much less frightening and frustrating to suction me than to assist me through a bad coughing spell.

As any caregiver knows, the burden of care can't simply be measured in hours. The stress and emotional toll can be much worse. For example, I can never be left home alone. Actually, it was probably every bit as dangerous for me to be alone for the last several years, but it is "officially" dangerous now! Thanks to having a live-in helper who, although she has a full-time job, allows my husband to get away in the evenings and on weekends, being available is now only annoyingly restrictive to him. And with two wonderful friends and my daughter, who were also trained in the basics of vent care, he gets time to run errands on weekdays and even take a full day off once in a while.

Being on the vent has made many things easier, however. Even though we have to pack a laundry basket of "just in case" equipment to leave home, I am a lot more portable and able to go places than before simply because I feel good again. For him, the good news is that I can accompany him most places. The bad news is that I can accompany him most places!

### *Clear Sailing*

I am no Pollyanna. I am well aware that my safe harbor is a temporary shelter. My ALS will continue to progress, taking the wind out of my sails and warping my timbers. There will come a day when I will need to decide if it is time to abandon ship, but in the meantime I can and do enjoy my life. It is so much easier to focus on the positives when breathing isn't a moment-to-moment battle! Thanks to the vent, today I am enjoying holding our newborn granddaughter, scanning the pages of children's books so I will be able to read to our 14-month-old granddaughter, working on Web sites. For years I was slowly disengaging from life, withdrawing, preparing to leave. Now I am re-engaging, rebuilding, and truly enjoying life.

It is a beautiful spring day and the breeze is gentle. I am going to weigh anchor, raise the mizzen mast and go for a little sail around the yard to make plans for this summer's gardens ... and I'll be planting perennials, not annuals. Life is very good here in Safe Harbor!

*Diane Huberty, 56, of Fort Wayne, received a diagnosis of ALS in 1985. She retired from her position as Neuroscience Clinical Educator at Parkview Hospital in 1995 and has since become involved in computer work including Web site design and photo editing and restoration.*

## *Remodel or Move?*

Don't sell, buy, build, or remodel until you can look ALS in the eye and deal with it! Plan for the day when you will be using a power wheelchair, not the manual chair that the ADA regs and contractors plan for. Plan for the day when you won't be able to stand to transfer from chair to bed or chair to toilet. Consider your rate of progression. That can be hard to predict, but a year into ALS you will know if it is fast progression. Two years in you will know if it is typical progression or slow. These things are emotionally tough to do, but the last thing you want is to find that all your work and expense was short sighted. Not all the modifications have to be made right away, but before you begin with any modification, have a good idea of what you will need down the line so moving or a major redo's won't be necessary.

Adding wheelchair ramps is usually the first on the "to do" list but this is exactly the kind of thinking to avoid. That doesn't mean that ramps should not be the first modification made. It means they shouldn't be done until you have looked ahead and made a plan to remodel or move that will address all future needs. This section is not arranged by what changes need to be done first, or even what changes are the most desirable. It is intended to go from room to room to come up with a complete plan.

Beginning with the structure of your current home (or wherever you plan to live as the ALS progresses), the first thought is whether it can work at all. Any home can work, but some, especially single story, are certainly more convenient and adaptable than others.

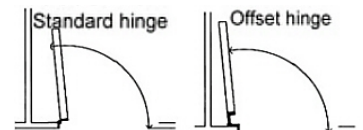
To evaluate the space, a tape measure is going to be needed. Two pieces of cardboard, one 4 x 4 feet to simulate a Hoyer type of patient lift, and another about 45 x 28 inches to simulate a power wheelchair, will be very helpful. The spec sheets on wheelchairs don't include the leg and foot rests so the length is more than the specs say! Same with the width. Armrests and joysticks add to the width of the chair so a chair is wider than the frame size given in the specs. 45 x 28 inches is close to the actual size of a power wheelchair-- unless you are going to need an extra wide chair. Lay the card board on the floor and "drive" it around the house to see if the doorways and hallways work.

So where do you start in planning for the future you don't even want to think about? Bathroom access is the first thing most people worry about but is a distant second when you look beyond the short term. Bathroom access, ramps, etc. are pointless if you can't get out of bed safely and conveniently, so, first look at the bedroom.

### *Bedroom*

If your bedrooms are all upstairs in a two-story home, staying upstairs after it becomes difficult to get you downstairs is not a workable plan. Getting you out for any event will become infrequent. In a Medical emergency it would be risky and in case of a fire very possibly fatal. In less drastic concerns, you would be isolated from day to day household events. Caregivers would be up and down the stairs repeatedly. There are ways to make the upstairs accessible but they are expensive and may not be workable in some home layouts. (More on this next.)

Regardless of whether an upstairs or downstairs bedroom space will be used, the room has to be accessible in a power wheelchair. An open space at least 5 foot square is needed to turn a power wheelchair around without making the turn in short back and forth moves. If the room is entered straight on, a 30 inch doorway will work. A 28 inch doorway can be widened about 2 inches by using offset hinges.



The most common problem is that the room is off a hallway and the wheelchair cannot make the turn to get through the doorway. A test drive with your power wheelchair cardboard pushed along the floor -- vroom, vroom -- will quickly show if a wider doorway will be needed. When widening a doorway, the light switch is generally close to the door and needs to be moved a few inches. That is an easy fix, but heating ducts or even plumbing pipes may have to be moved. That is more involved but usually possible. A replacement doorway 36 inches wide is generally manageable but going even wider using two bifold doors is a better guarantee that it will be wide enough. Pocket sliding doors are another possibility but require the full door width to be unobstructed space inside the adjacent wall.

The bedroom should be able to accommodate twin beds with at least one of them not pushed up against the wall. Couples can stay in a regular bed at first, but a hospital bed will be needed down the line. Separate beds may seem unnecessary, but there will come a day when a hospital bed is the best. Hospital beds have important features the caregiver will need.

- They are single beds that allow the caregiver to work from either side.
- The entire bed can be raised to a height that makes it easier to rise from sitting on the side of the bed to standing.
- The height also prevents back strain for a caregiver moving, turning, bathing the patient.

Usually a caregiver spouse is sleep deprived and sleeping in a separate room can help. Even though they will be up and down during the night, a separate room without the patients every sound can be better. Having the caregiver sleeping upstairs has them on the stairs when they are half asleep or out of hearing range if the call system fails (out of reach, dead batteries, not turned on) is too risky!

A critical consideration in a bedroom is the type of lift that will be used to transfer from bed to chair and back. The choices are between a Hoyer lift and an overhead lift.

The standard patient lift is frequently called a Hoyer lift. "Hoyer" is a brand name and there are many other brands. When planning for this type of lift space can be an issue. A Hoyer type of lift is about four feet long. The width is nearly that when the legs are opened out to prevent tipping while in use. Use your Hoyer size cardboard and see if you can fit and turn it on the floor in the hallways and rooms you will be working in.



Transfers from the bed to the lift to a wheelchair require space at the bedside for the lift, and space for the lift and wheelchair to be positioned for the transfer. Since a power wheelchair is also about four feet long it will take a lot of space for this -- and more if you want the lift and wheelchair out of the way between uses. It will be possible to roll the patient in the lift out of the bedroom for the transfer, so that can reduce the space needed in the bedroom. A lift will also require space when not in use so that also needs to be considered in the available space.

A Hoyer or similar lift is hard to push and turn on carpeted floors. A change from carpet to solid flooring will make it much easier.

Overhead lifts are a good choice for patient transfers, especially when space is an issue. Transfers are easier and faster than with a Hoyer type because they take the patient directly from bed to wheelchair or commode. There are three types of overhead lifts available.



Ceiling mounted lifts do not take any floor space and can be installed flush with the ceiling. They may be straight, curved, and tracked to go from room to room. Construction work to strengthen or support ceiling joists may be necessary.

Free Standing Lifts can be self-installed with no attachments to walls or ceiling needed. Quickly dismantle to move.

Wall Mount Lifts mount to the wall along side a toilet, bathtub, or bed so don't take floor space. Require reinforcement of wall stud.



In addition to space and flooring requirements, there are other things to consider about Hoyer type of lifts. See Patient Lifts, Page 95.

When you start adding up all the electrical equipment that is likely to be used in the bedroom, it is surprising: lights, electric hospital bed, suction machine, nebulizer, electric blanket, fan, TV, CD Player, call system, electric or battery powered lift, and charging outlets for the vent, wheelchair, and every other battery operated thing that requires recharging. (Label every battery charger and cable with the name of the piece of equipment it goes with or you will end up in Charger Hell.) Thankfully not everything will be in use at once, but check what other rooms are on that circuit. Living rooms can be electricity hogs too. TVs, sound systems, computers and all their peripherals, lighting, space heaters and fans. Kitchens definitely would not be good companions on a circuit with either your bedroom or living area! Few houses have enough outlets in the right places for what you need to plug in so it is very helpful to add outlets or power strips. Having the power supply throughout the house evaluated is a good idea. The most that is likely to be necessary is adding a circuit to reduce the load on another circuit or circuits.

### *Stairs*

A common misconception is that a stair lift will get you up and down the stairs in a two story house even as ALS progresses, so let's discuss stair solutions for two story homes. A chair lift in the stairway may work for some time, but there will come a time when a person with ALS (PALS) cannot sit comfortably and safely on the small stair lift chair. Neck and trunk weakness will leave you a rag doll in the chair which is not designed with trunk support, a head rest, recline, or a foot rest large enough to keep your feet in place.

Although a chair lift can easily be configured for stairway with landings and turns, space at the top and bottom of the stairs is just as important. If it can be configured to allow plenty of space at the bottom to bring a power wheelchair up to, or ideally, along side the chair lift seat, transferring to the chair lift will be easier and extend the time the lift can be





used. Otherwise, the usefulness of the chair lift ends when walking with assistance from the wheelchair to the chair lift is no longer possible. At the top of the stairs, another wheelchair or chair on wheels (a commode chair with arms and adjustable height works well) is needed to get to the bedroom and bathroom. Again, having room to transfer is important. Depending on ALS progression, this can work well, but when standing to transfer becomes difficult, all these transfers are very hard on caregivers and may not be possible. A Hoyer lift can do the job but again space is an issue -- and you will need one downstairs and another up.



Another possible solution is a wheelchair stair lift. Similar to the wheelchair lifts used in vans, they move the wheelchair and passenger up and down a flight of stairs. Many brands are available and some will fit a staircase less than 36" wide. They do require more space at the top and bottom of the stairs for loading the wheelchair but some brands offer the ability to curve to the outside of the stairway for loading. The platform has edges that fold up to prevent rolling off, and the platform folds up to allow normal use of the stairs. The weight limit for these is generally 600 to 650

pounds. The weight of power wheelchair can be anywhere from about 200 pounds to 400 or more. That can make it unusable for some.

An elevator is a great solution but expensive, though probably cheaper and less disruptive than moving. More importantly, an elevator is a solution that increasing weakness won't make unusable. Finding space for an elevator can be difficult since it has to be large enough for a power wheelchair and an attendant. Backup power for the elevator as well as inside the elevator is necessary. The IRS considers elevators (but not stairway chair lifts) as adding to home value so none of the expense qualifies as deductible.

Similar to an elevator, an vertical wheelchair lift may work. With the same configuration as a wheelchair lift in a van, but possibly without room for an attendant to ride with you, it may be possible to fit it in a smaller space than an elevator. It does not have to be placed



along a stairway so it can lift directly into an upstairs bedroom. Although many vertical lifts are limited to lifting 6 feet or less, there are others that can lift a full story or more. It can be left open or enclosed (which may be required). Because it isn't mounted at an angle to a wall, weight of the passenger and chair is less an issue.



## *Bathroom*

Bathroom concerns are high on the list of home modification needs. A hard look at the reality of your situation is important. How fast is the ALS progressing? A big, beautiful spa bathroom is wonderful but how long will the PALS use it? All too often the progression of ALS leaves only a few months where the new or remodeled bathroom is used. Is there money readily available for a big bathroom project or will your family be paying off a loan after you are gone? Feeling that a big bathroom project is essential may be just a manifestation of denial of the reality of progression and life expectancy.

The resale value of a home will go up with the addition of any bathroom, but an accessible and handicapped equipped bathroom may not be a big selling point. If the bathroom is located off a bedroom it will be a plus. If it is awkwardly placed off a living room or family room, steals significant space from another room, or turns a three bedroom home to a two bedroom, the resale goes down. Buyers should consider that they are likely to need it down the line, but they don't!

## *Showering*

Some PALS have been able to shower up until the end, but for most showers get further apart as bed baths become all that is tolerated. Showering a wheelchair person requires extra transfers and is far more work and much more time consuming than a good scrubbing while sitting on the toilet. A bed bath is even less exhausting for the patient, especially when breathing problems begin. No one needs a daily bath. A wash of pits and bottom and a shampoo every other day will do fine between weekly full body wash ups. A good soak in a shower would feel great but unless your progression is proving to be very slow, a bathroom remodel or addition price tag isn't justifiable if cost is at all a consideration.

Whatever the plan, at least some bathroom remodeling will likely be needed. When there is no downstairs bathroom, the best option is to add one but this may not be possible for financial or space reasons. No bathroom on the main floor will mean a lot of trips upstairs for the caregiver to empty urinals, bedpans and commodes, but all other bathroom activities -- bathing, shampooing, etc. can be done in the kitchen or in bed. A half bath may exist or be possible to add and may possibly be made accessible for a wheelchair. Even if it is not accessible to the wheelchair, it will be a timesaver for caregivers.

### *Space and Layout Considerations*

When figuring out the space needed and layout for the bathroom, I suggest drawing it out full scale (including toilet, sink, etc. and any doorways and hallways the chair must navigate) in chalk on your driveway and do some wheelchair and transfer simulations. What looks good on paper doesn't always work as expected -- even if designed by a contractor! Because the room must be set up for the eventual power wheelchair, plan for a wheelchair length of at least 45 inches to include the footrests. For testing, your power chair sized of cardboard can be moved across the layout to check the space.

If the problem with an existing bathroom is mainly a lack of floor space to maneuver, you might consider replacing one wall of the bathroom with a set of large (6 ft total) double folding doors. That opens the bathroom up to give a lot of "elbow room" when needed and yet allows the space to be returned to its normal use at other times.

It may be possible to convert a tiny half bath into an accessible space. Double doors can turn a hallway into part of the bathroom for wheelchair maneuvering -- and back to a hallway when the doors are closed. See *An Accessible Half Bath*, Page 31, to see how we did it!

Allow plenty of space for your assistant to stand squarely in front of you and take a step or two backward as he/she lifts you. We were surprised at how much space that took -- even though we had plenty of experience with narrow bathrooms where John ended up plastered against the wall and unable to maneuver once he had me up on my wobbly legs!

Even when I used a sliding bench to slide sideways into the shower, it was not worth the effort! I ended up exhausted and shivering. My husband/caregiver was wet and sweating. I gave it up in favor of a wash up while sitting on the toilet and a shampoo at the sink which we modified to a roll under vanity. When transferring to the toilet later required a lift, we switched to a simple bed bath of my lower half. One soapy wash cloth, one wet, and a hand towel. No basin of water to spill! Dress my lower half and use the overhead lift to get me into my wheelchair. Off to the bathroom for the rest of my bath with me at the roll under sink. Shampoo, tooth brushing are so easy there! Finish with dressing my top half. I am clean and dressed in half the time and with no lifting and neither of us is exhausted.

Have the entire toilet moved away from the wall behind it. A power chair cannot be backed up to the wall enough to be side by side or even positioned at a good angle with the toilet if it is against the wall.

#### *Fixtures and Gadgets*

- A roll under sink is worth the cost, even essential, but a roll in shower isn't. Need for a full bath with a roll in shower is debatable in any remodel and can actually be done without entirely.
- Choose sinks with the narrowest rim and have them positioned as close to the front edge of the counter as possible. When you can't spit with any force, that will be helpful!
- Price Pfister makes a lavatory faucet with a pull out sprayer that makes shampooing easy.
- An elevated toilet helps tremendously with transfers and is well worth the cost. (See toilet options here.)
- Waterproof flooring, of course.
- Grab bars
- Install a floorboard or ceiling heater.

Eventually standing transfers won't be possible and some type of lift will be necessary. Refer to Patient Lifts to see the types available. Few bathrooms are big enough to maneuver a Hoyer lift in so an overhead lift is better and far less expensive than enlarging or adding on a bigger bathroom. Even though an overhead lift won't be needed right away, have the necessary ceiling joist supports or wall studs put in if any work in the bathroom is done.

If the cost of overhead lifts in the bathroom and over the bed is out of the budget, a Hoyer lift is easier to use in the bedroom than the bathroom. If it comes down to just one lift, choose the bedroom. A lift there will be needed to get in and out of bed and can be used to transfer to a commode. Yes, a commode seems unthinkable early on, and that is the type of thing I meant by looking the long term/advanced stage of ALS in the eye and dealing with it. Anyway, when you reach the point of needing to be lifted on and off the toilet and having someone wipe your bottom, modesty and squeamishness are mitigated by simple practicality.

Medicare does pay for bathroom equipment related to toileting. If you file for a commode it is covered even if it has a full back and headrest and wheels. Just make certain it has a cut out seat for use as a commode when wheeled over a toilet. And DO NOT refer to it as a "shower chair" or to needing it for that reason. You do need a commode on wheels with back and head support for toileting and that is how it should be filed. An occupational therapist from your MDA/ALS clinic could help assure that it is filed correctly if paying for it yourself isn't possible.

#### *Ramps*

Now that you have evaluated your home for its indoor space, look at the lot. The levelness of the lot goes into determining how long a wheelchair ramp would have to be. The requirement for a building permit for a ramp is that the ramp be 1 foot long for every inch it has to climb. A house with a slope or steps to the doorway can result in ramp that simply won't fit on the property. A ramp that has to have a turning landing adds considerably to the cost. For safety, make sure that there are two wheelchair accessible exits from the house.

ADA regulations for the construction of wheelchair ramps do not apply to private homes unless that home also houses some type of business such as a baby sitting service or home based business. State, city, and county building codes will apply and they may be based on ADA regulations as well

as having other regulations such as distance from lot lines, etc. So the place to start is to investigate any regulations that will apply to your building permit. Even though ADA regs don't specifically apply to your home, take the time to review them for information on space and safety features you may not have considered.

Do you need a building permit? Unless you live in an area where building codes don't exist, yes, if the ramp is going to be attached to your home. Getting by without a permit is certainly possible, but if anyone notices your little construction of project is lacking a permit notice, they may rat you out to the building board. Why would anyone object to a wheelchair ramp? Aside from pure meanness from the neighborhood grouch, some may think that a ramp on your house will reduce the property value of their home. Others may be sticklers for the rules or ticked off that they had to get a permit for something they did. Re-doing a ramp halfway through construction is not fun, so unless your home is pretty much out of sight, a permit can save trouble. Of course, meeting codes, dealing with inspectors, and waiting on inspection visits present its own set of hassles, but local codes are more likely to allow you reasonable exceptions to fit the ramp to your lot.

The biggest hurdle to going with ADA regs is the length of a ramp. They require a foot of length for every inch that the ramp has to climb. A ramp up to door or landing that has three or four steps can be as much as 28 feet. If the lot is sloped, that adds to the length. You can easily skimp on the length and still be able to get a wheelchair or power chair safely up and down, but you have to consider the strength of the person who will be pushing you in a manual chair as well as the power of a power wheelchair to climb a shorter ramp. Power wheelchair websites generally include the slope a particular chair can handle. Regardless of the fact that the one foot long for every inch of rise seems excessive, and may well be, building codes probably prohibit any less. A contractor is not allowed to build a ramp that doesn't meet local building codes. Thankfully, the building permit inspector can grant a variance and shave the foot per inch requirement somewhat if the ramp simply won't fit on your lot otherwise.



If you have a high porch, deck, or front door landing, your first mental image of a ramp may be a long, long ramp or one that zigzags like the barricaded line for the most popular ride at the amusement park. A much better solution would be an outdoor vertical wheelchair lift which can be positioned to lift



you onto a porch or deck. The only modification needed may be expansion of the porch, deck, or landing to make room for the wheelchair to turn to the door.

Pre-built ramps are available and save time and effort. For a front door ramp, my choice would probably be for having a ramp built. That would give you more choices about how the end result would look, could be landscaped, etc. Depending on the height of the porch, a ramp that meets ADA and city regulations can be really ugly if not well laid out. But if curb appeal doesn't matter to you, a pre-built one would be easier and probably less expensive. Ask around though, there may be volunteers or organizations who will do the work for free if you pay for the materials.

To me, a ramp in the garage is definitely the way to go even if it means leaving a vehicle outside. If your wheelchair van fits in the garage, it is wonderful to be able to load and unload out of the wind, rain, snow, heat and cold. And even if it doesn't, it is still nice to be able to zip in and out of the garage rather than waiting outside. The ramp is also out of sight and doesn't mess up the looks of your house. Some outdoor carpet on the ramp will give it traction, reduce dirty wheel tracks inside, and, being inside, you don't have to worry about ice or snow on it. (And unless you have nosy, nasty neighbors, it can be built without anyone noticing and sending the building inspector after you.)

You can avoid the whole permit/inspection thing if the ramp is not attached to the house and could be considered temporary and portable. Simple aluminum ramps come in many lengths. This is a good option for some houses but not all. A portable ramp would be a good idea for a second exit in case of fire. That ramp could be shorter since it wouldn't be inspected. As far as building material, outdoors use treated lumber just as used for deck building. Anyone with basic building skills should be able to do the job with less than 20 trips to Home Depot!

Often the garage floor is just an inch or two short of level with the driveway. A bag of cement or asphalt can easily be shaped into a short ramp. Asphalt doesn't have to be mixed but cement will hold up longer. The same can be done at the threshold from garage to house or side walk or porch to a door.

### *Kitchen*

I hesitate to say something so negative, but even if you enjoy cooking, major kitchen remodeling probably is not worth the cost, especially if you are already experiencing much arm weakness. Most adaptive aids for cooking are helpful in replacing hand strength and grip but cannot compensate for the loss of arm strength. Another concern is that working with hot foods presents big safety risks.

There are several things you can do inexpensively to allow you to continue to do at least some of the cooking as long as your arms hold out:

- Put slide out trays in all the bottom cabinets and store all frequently used items there.
- Use a stool on wheels to scoot around the kitchen but still be up at countertop height. Chairs made especially for this are available from some adaptive aids web sites.
- Place the microwave on a cabinet that is deep enough to allow you to slide dishes out of the microwave on to the countertop rather than having to carry them to another counter.
- Remove the doors and floor of the cabinet under the kitchen sink so that you can get your knees under it. (If there is a possibility your knees will bump the pipes, cover them with insulation tubing to prevent burns.)
- If you can afford it, you might consider a countertop range, again with knee space under it. (It is a trade-off when you start making knee space out of your lower cabinets -- you lose the only storage space that you can easily reach!)

### *Flooring*

When you do an internet search on the best flooring for wheelchair use, you get conflicting answers. Most answers to the question are focused on manual wheelchair use so they will say that carpet should be removed. It is much harder to propel a manual chair on carpet, but it makes little or no difference with a power wheelchair! Only areas where a Hoyer lift or wheeled commode will be used need solid flooring. If you want to keep the carpet in those areas you can put plywood or office chair mats (solid ones, not the flexible kind) on top of the carpet where needed.

The next question about carpet is whether a power wheelchair will ruin it. That pretty much depends on the quality of the carpet and how well it has been laid. Even a medium quality carpet should not bunch up from wheelchair traffic if it was well laid. The exception might be if the carpet was spliced together at a spot where the wheelchair will be making tight turns. As with foot traffic, a wheelchair will mat down paths but doesn't seem to ruin the carpet any faster than foot traffic areas.

The biggest problem with power wheelchairs and carpet is that the wheels track in rain, snow, mud, sand, and plain old dirt. A simple floor mat at the door is worthless. It takes a long carpet runner to even begin to dry the wheels, and they will still carry stuff in the treads. There are a few wheelchair tire cleaning devices for power wheelchair users who are off road enough to warrant buying one, but for most of us, the best solution is a friend with a towel and possibly a brush. And not visiting people with white carpet!

Hardwood floors are often recommended because any wear from the wheelchair can be fixed with sanding and refinishing the wood. (We have so little hardwood forest left, and if you look into buying hardwood flooring you will see that it is Brazilian hardwood. The idea of destroying forests in other countries just to support the fad of hardwood floors is repugnant. Bamboo is very strong and is environmentally far better and can be engineered to look like other hardwoods. End of ecology rant!)

Good quality wood laminate will stand up to a wheelchair but tends to be noisier and doesn't add to home value the way the snob appeal of hardwood does.

Tile floors are durable and easy to clean. Profession installation will assure that the subfloor is level. Good quality tile is a must to prevent cracking under the weight of a power wheelchair.

Sheet vinyl is not necessarily the most economical option because the underlayment often needs to be replaced and the flooring installed by professionals or any guarantee will be void. But medium to high-quality vinyl will hold up to a wheelchair and is the easiest flooring to maintain. Later, new flooring can be laid over it without having to remove it.

Vinyl tiles offer the same easy care and can be installed by yourself. They would be fine for a few years of power chair use but might begin to peel up in time.

Vinyl plank wood-look flooring is an easy do-it-yourself project. The planks lock together like hardwood or laminate. They are quiet, can be 100% waterproof, and wear as well as comparable quality sheet vinyl. The planks may tend to pull apart at the ends of the planks where the wheelchair moves over. This doesn't happen on concrete floors and may be less a problem with higher quality/thickness planks. Of course, a very solid underlayment for wood floors is necessary. If some planks either do begin to gap, they can be kicked back in place by wearing sneakers.

### *General Suggestions*

- A living room layout that will accommodate an accessible desk -- your Command Center -- will keep you in on family activity and hopefully give you a view of the outdoors.
- Move frequently used light switches and outlets to the level of wheelchair arm rests. Make the light switches the larger easy to press switches.
- Change door knobs to lever style handles.

### *Tax Deductions*

A large expense in ALS is making your home accessible. Changes to your home to make it accessible to the taxpayer, the taxpayer's spouse, or dependent who lives there are deductible as Medical Deductions. Large projects such as adding a bedroom, bathroom, or elevator can be partially deductible. If the cost of materials and labor is more than the added value to your house, you can deduct the difference. The IRS has a list of home changes that it does not consider as adding to the value of your home and are therefore entirely deductible. They include:

- Wheelchair ramps inside or outside, including landscape grading if needed
- Widening or modifying exterior or interior doorways or hallways

- Installing railing or support bars in bathrooms, and other modifications intended to increase bathroom safety
- Lowering kitchen cabinets and equipment, and other kitchen modifications
- Changing the location of or otherwise modifying electrical outlets and fixtures
- Porch lifts or stair lifts
- Modifying stairs
- Adding handrails or grab bars, including in areas other than bathrooms
- Modifying door hardware

If you "upgrade" as part of these changes, only the basic change is deductible. Example: Replacing flooring may be necessary when widening a hallway. Upgrading from vinyl flooring to ceramic tile would be deductible only to the amount that new vinyl flooring would have cost because tile would add to the value of your home.

To defend your claim if the IRS challenges it, really big projects should include good record keeping of costs. Written advice from your physician or physician ordered therapist's evaluation about the need for the improvement to your home, and before and after appraisals to show any increase in value due to the improvement would be helpful as well.



## Equipment Needs

### *Call Systems for ALS Patients*

A major concern for ALS patients and their caregivers is finding a reliable method for the person with ALS to alert the caregiver that help is needed, especially when hand strength is weak and speech hard to understand. In that situation the best equipment uses a capability switch to trigger a chime or other sound. This requires equipment that is switch adapted.

A capability (ability, disability) switch is an easy to press button that attaches to equipment through a jack and cable. Capability switches are available from several online sites. Many configurations allow the best choice for whatever movement is still available in any part of the body as well as wheelchair or bed placement. A few examples:



A disability cell phone would seem to be a great solution, and some iPhones can be used with a capability switch but require speaking the name or number to be called so their use is limited with ALS. You also have to have the phone at eye level to see the menus and select the number to be called. It isn't clear if the phone can be set to automatically dial one specific number when the capability switch is pressed, rather than go through the menus.

Some people use a baby monitor. It doesn't require any hand strength but does require a fairly strong voice or ability to make some sound, so its use is also limited.

The most frequently used call button is a battery powered doorbell. The patient has the doorbell button and the caregiver has the "ding-dong" chime sounder. This works well -- until the button becomes too hard to press. Popsicle sticks taped over the button can turn it from finger control to a hand grasp, but this also becomes a problem as weakness progresses. In the past it was easy to adapt a doorbell to use a capability switch instead of the doorbell button to trigger the doorbell chime. Today's doorbell kits use lithium coin sized batteries which can't be adapted to use a capability switch.

There are few disability equipment providers that adapt their call systems for capability switch use! However, you can find a call system with a transmitter that is battery powered by A, AAA, C, or D batteries, adapting it yourself is easy. (It doesn't matter what batteries the receiver uses.) Instructions are included for the call systems I have included here.

### *Options*

I have searched the Internet for hours, looking for call systems that are already capability switch adapted or have the right batteries to allow them to be converted to a capability switch. I am showing only wireless models that the caregiver can carry with them and one that can use extra alarms in other rooms. Alarms that only sound in the room with the person with ALS don't allow the caregiver much freedom to move around the house. Note that none of the listed call systems include the capability switch in the listed price and none of the pictures show the capability switch attached to the transmitter.



### Vibrating Personal Pager

\$61.95

<https://www.adaptivetechsolutions.com/vibrating-personal-pager-switch-adapted/>



#### Pros

- Adapted for capability so set up is easy.
- No monthly charge for service.
- 100 ft. range indoors.
- Vibrates or chimes
- Requires 1 12-volt battery (included).
- Receiver has a belt clip and table stand.
- Transmitter has a lanyard

#### Cons

- Batteries can run down and cause it to fail.
- Requires 2 "AAA" batteries (not included)
- Capability switch not included.

### Wireless Attendant Call Button Vibrating Alert Chime

\$49.48

<https://www.adaptivetechsolutions.com/pd-wireless-attendant-call-button-disabled.cfm/>



#### Pros

- Already adapted for a capability switch so adding one is easy.
- No monthly charge for service.
- Large button on the patient's transmitter.
- Belt clip on the caregiver's receiver.
- 500 foot range to allow caregiver to be nearby outside.
- Variety of chime sounds with adjustable volume.
- Vibration setting allows a caregiver to be alerted while in a noisy environment or if hard of hearing.
- Requires 1 12-volt battery (included).

#### Cons

- Batteries can run down and cause it to fail.
- Requires 2 "AAA" batteries (not included)
- Capability switch not included.

Wireless Attendant Call Button Vibrating Alert Chime & Plug  
In Receiver  
\$59.48

<https://www.adaptivetechsolutions.com/wireless-attendant-call-button-vibrating-alert-chime-with-plug-in-chime/>

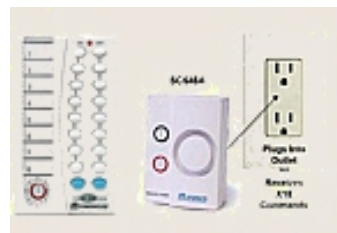
Identical to the above system but includes a wall plug includes an A/C plug in receiver chime for indoor use, as well as the battery operated portable receiver chime.



X-10 Plug in Doorbell Call System  
\$34 - \$45

<https://www.x10.com/x10-home-automation.html>

<http://www.smarthomeusa.com/brands/X10.html?page=2&sort=featured>



Pros

- No monthly charge for service.
- Caregiver doesn't have to carry the chime.
- The chime can be plugged into an outlet in any room.
- Additional chimes can be plugged into outlets in other rooms, patio or porch.

Cons

- Set up requires adding a connection for a capability switch.(Instructions follow.)
- Chime only, no vibrate.
- Batteries can run down and cause it to fail.
- Will not work in a power outage.
- Capability switch not included.

*Added Range Call Systems*

Increasing the range of a call system has definite advantages for reasonably stable patients. The caregiver can be free to go outside to do lawn work, gardening, walk the dog, visit with neighbors. Neighbors can take the pager and stay at home while being available quickly. The likelihood of the need for an immediate response may keep the caregiver within a minute or two from home, but many PALS, even those on a ventilator, can rely on a longer range call system. A longer distance comes at a higher price, however, because it requires a true paging system. These are the ones used by restaurants, hospitals, and other places where customers may have to wait a long time and a pager allows them to leave the waiting area.

LRS manufactures many types of paging systems with no monthly charge for service. The LRS website offers very little about the various systems, and you have to request prices. Go to foodsoftware.com instead for complete information, prices, ordering, and support.

PT01 Pager System

- Transmitter and pager \$149.95
- Up to 1,000 feet (1/5mile)
- Large button

Butler XP Pager System

- Transmitter and pager \$199.95
- Up to 1,320 feet (1/4 mile)
- Smaller button

Any transmitter's range is reduced by being in or near concrete or steel structures, steel and sheet metal

<ul style="list-style-type: none"> <li>• Beep or vibrate</li> <li>• Low battery indicator</li> <li>• Belt clip</li> </ul>	<ul style="list-style-type: none"> <li>• Beep or vibrate</li> <li>• Low battery indicator</li> <li>• Belt clip</li> <li>• Higher price due to additional options not useful for ALS</li> </ul>	<p>cabinets, shelves, etc. For the best range, place transmitter in front of a window. (Extension jack cables are available in many lengths.) These transmitters can send only one pre-programmed message but even that isn't necessary. Who else would be paging your caregiver?</p>
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*To convert for use with a capability switch you need:*

For X-10	For Pagers
<ul style="list-style-type: none"> <li>• PalmPad Remote Control HR12A (\$15-\$20). This remote control is overkill for the single chime we need, but X-10's smaller remotes use lithium batteries which won't work with a capability switch.</li> <li>• 4 AAA batteries for the PalmPad Remote Control.</li> <li>• Battery Interrupter (\$13 at Adaptive Tech)</li> <li>• A battery interrupter is a small copper disc wired to a female jack. Battery interrupters come in two sizes, small for AA and AAA battery devices and larger for C and D battery devices. This remote control requires the smaller size.</li> <li>• Remote ChimeSC546A (\$19-\$25) The chime plugs directly into any outlet. Extra chime units can be added in more rooms.</li> <li>• Capability switch.</li> <li>• 3 sidedTriangle file.</li> </ul>	<ul style="list-style-type: none"> <li>• ARCT PT01 Pager System or Butler XP Pager System</li> <li>• Batteries, 2 AA, and 1 AAA.</li> <li>• Battery Interrupter (\$13 at Adaptive Tech) A battery interrupter is a small copper disc wired to a female jack. Battery interrupters come in two sizes, small for AA and AAA battery devices and larger for C and D battery devices. This remote control requires the smaller size.</li> <li>• Capability switch.</li> <li>• 3 sided triangle file</li> </ul>

This Video from Enabling Devices shows the easy assembly steps.

<https://www.youtube.com/watch?v=N3P8jv6wufA>

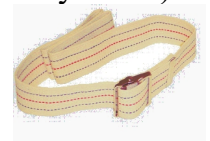
Open the battery compartment of the Remote Control/Transmitter and place the disc of the battery interrupter between a battery and its contact point. (Take the battery out and put it back in place as you insert the battery interrupter.) If the disc is too large for the compartment, use scissors or nippers on the edges to shape it to fit. Use the three-cornered file to cut a groove in the battery compartment lid for the battery interrupter wire so that the compartment can be closed.

<ul style="list-style-type: none"> <li>• Press one of the buttons on the PalmPad Remote control to ON.</li> <li>• The battery interrupter has a 1/8" plug on the end. Insert the 1/8" jack of the capability switch into it.</li> <li>• Set the number dial on both the PalmPad Remote Control and the Remote Chime to the same number.</li> <li>• Plug the Remote Chime into any outlet. Press the capability switch to make the Chime sound.</li> <li>• To add chimes in other rooms, set the number to match and plug into a wall outlet. If interference causes chiming without pressing the capability switch, try changing the numbers on the remote and the chimes to another set of matching numbers.</li> </ul>	<ul style="list-style-type: none"> <li>• The battery interrupter has a 1/8" plug on the end. Insert the 1/8" jack of the capability switch into it.</li> </ul>
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### *Patient Lifts*

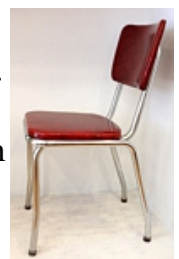
Before discussing the lifts available, let's start with the moment many of us finally get serious about getting equipment to lift us up. That moment is when we end up on the floor and no one can get us up by themselves. There are ways to handle this short of calling 911. (Though I found this a delightful solution because I got 4 hunky EMS and Firemen to lift me and dance me to my chair.)

Lifting a person who can help to some extent can be made easier using a gait belt. With the belt around the waist it provides a good place to grab and lift rather than pulling on an arm, clothing, or using a bear hug.



Another way to make the task easier is to place a low footstool in reach, then lift the person to a sitting position and put the footstool behind him or her. Using a gait belt or a bear hug from behind and whatever arm strength the person has to boost himself, move up onto the footstool. Repeat the process moving to a chair or to a higher stool and then a chair.

Place a simple kitchen chair next to someone unable to get up from the floor. The best chair is one with a flat back. A high back is ideal for someone with neck weakness. A chair with a curved back is harder to slide onto and knobby spindles are uncomfortable. After checking for broken arms, legs, or shoulder, hip or neck pain, turn the person on his back. Place the chair on its back along side of him. Lifting with your legs so you don't hurt your back, slide and lift him on to the chair so he is in a sitting position but on his back on the chair. A strong person can then lift the chair upright. If the fallen person is "floppy" you will need an extra person to keep him from falling off the chair.



### *Toilet Lifts*

One of the first places we need lifting help and don't want to ask for it is in the bathroom. Toilet rails attach easily and securely to the toilet and are very helpful when legs are weak but arms are strong.



- ◆ Toilet Seat Risers fit on the top of standard or handicapped toilets to give an average of four inches extra height. Other heights are available. They work well as long as

standing is safe. Later, when assistance to remain standing, walk, or transfer is needed, they reduce back strain for the caregiver.

- Inexpensive, available at drug stores or any place selling patient care items.
- Most are easy to install. May be easy enough to remove for single person use and portability.
- The easier they are to remove the less stable they are likely to be.
- Those with grab bars must be clamped tightly and securely to the toilet.
- Require frequent cleaning.
- Range from obvious to downright ugly.



#### ◆ Toilet Risers

- Fit beneath the toilet and raise it 3 to 4 inches.
- Can be used under standard height toilets or handicapped height.
- Available for round (less than \$100) or elongated (\$200) toilets.
- More sanitary than seat top risers.
- Not obvious to visitors. Can be painted to match fixtures.
- Requires very basic plumbing skills to install.



#### ◆ Power Toilet Lift

- Can lift about a foot up for easy standing and maximum independence.
- Are expensive for short term use.
- Will not work with all bidet systems. (Bidets wash your bottom and delay the need for assistance wiping.)



### *Transfer Lifts*

#### ◆ Hoyer Lifts

The standard patient lift is frequently called a Hoyer lift. "Hoyer" is a brand name and there are many other brands. Compare prices for used equipment on Craig's List. It is often minimally used and cheaper than buying new even if you have insurance/Medicare to cover a new one. Because you can buy locally, you can try out the equipment and don't have to pay shipping.

The brand of lift you buy makes little difference but the type of sling you use with the lift is critical to how it will work for you! The sling should be purchased separately from the lift in order to get the right type. Almost any sling will work on any lift.



A hygiene sling, also called a split leg sling or U-Sling is by far the best. These can be put under the patient or removed while in a chair or in bed. Standard "commode slings" are a major effort to get under the patient and can't be adjusted as well to get the person sitting more upright. Look for the simplest, uncluttered design and get one with a headrest. In my experience, sling sizes tend to run large so downsize if the ranges are close. Weight loss in ALS adds to the possibility of oversizing.

First attempts at using the lift and sling are seldom successful! Have extra help and don't use the patient as the test crash dummy! With patience and adjustments, only one caregiver will be needed for a smooth lift. Refer to Toileting Using a Lift for information on how to use the lift and sling.

A lift becomes a necessity at some point, but I have found that an overhead or ceiling mount lift over the bed is much more convenient than the Hoyer so you will want to consider that option before buying.

Things to consider when buying a Hoyer type of lift:

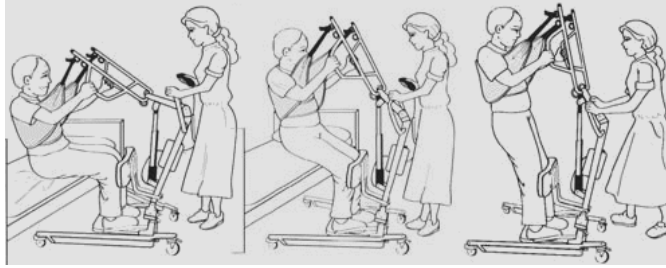
- Can it be lowered to the floor to pick up a person who has fallen?
- Will it fit under the bed you will be using? Most require 4 to 7 inches of space.
- When the base is spread for stability, will it fit around your recliner or wheelchair?
- Will it fit in your bathroom and bedroom? Requires at least a 5 foot square turning area. Suggestion: Before buying get the specifications list. Find the full length and width (with the base opened out). Cut cardboard to this size and see if you can fit and turn it on the floor in the hallways and rooms you will be working in. You may find that the best way to use the lift is to move over a commode, not the bathroom toilet. That is not ideal and definitely an emotional adjustment, but is far less expensive than a bathroom remodel.
- Weighs about 100 pounds. Consider the weight of the person and the ability of the caregiver to push the total weight.
- Hard to push and turn on carpeted floors.
- Requires storage space.
- Does not work as a bath lift for standard tub.
- Manual (hydraulic \$550 and up. A battery powered (\$1,300 and up) is easiest for caregivers.
- The bar (cradle or carriage) that the sling attaches to may have two, four, or six hooks. Two hooks are all that is needed. A big cradle is heavy, awkward and not fun to get hit on the head with!
- Some portable lifts are available but may not be usable with other brands of slings and the sling may not have head support.

#### ◆ Stand Up Lifts

- Stand Up Lifts have a sling backrest behind you and a padded knee block in front to keep your knees from buckling.
- Most have an optional 2nd "buttocks sling" that wraps under the thighs to allow the person to sit during lifts and while being moved to another room.
- Require some weight bearing strength and some upper body strength and stability.
- Most require the arm strength to hold on to a bar.



- Available in manual or electric (battery) power.
- Cannot be lowered to the floor to pick up a person who has fallen.
- One brand, the Easy Pivot, lifts you to stand but leans you forward over a cushion so upper body strength isn't needed. It may interfere with breathing however.

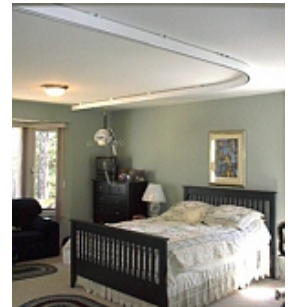


#### ◆ Overhead Lifts

There are three types of overhead lifts available:

##### 1. Ceiling mounted lifts:

- Are the most expensive.
- Can fit any space, can turn or curve.
- Can be installed flush with ceiling, on ceiling, or hung from ceiling.
- Does not take any floor space so are the least noticeable in a room.
- May be tracked to go from room to room.
- Requires company installation for safe use.
- May require construction work to strengthen or support ceiling joists.



##### 2. Free Standing Lifts:

- Lower cost than ceiling lift.
- Self installation.
- Over the bathtub frame option.
- No attachments to walls or ceiling so no repairs when removed.
- Easily and quickly dismantled to move or take along.



##### 3. Wall Mount Lifts

- Mounts to wall along side a toilet, bathtub, or bed.
- Compact and space saving.
- Attached to wall so it is not portable.
- May require reinforcement of wall stud.
- Swing bar must be positioned for a specific touch down spot.



## *Wheelchairs*

Putting aside the unanswered questions of "Use it or lose it" versus "Exercise hastens progression", there comes a point when refusing a wheelchair is not only impractical but unsafe and self-defeating. Using a wheelchair doesn't mean giving up walking completely and it doesn't mean giving in to the disease. Letting the disease limit your life too much, too soon, and cause dangerous falls is giving in to ALS! If your subconscious is fighting a wheelchair because it is giving in to ALS, your subconscious is dead wrong. It isn't fighting the disease, it is trying to wish it away. Fighting ALS is all about finding solutions, not letting the disease rob you of things you could still do. We often hear the phrase "confined to a wheelchair." That is so wrong! It should be "freed by a wheelchair". Freed to move around independently, freed from exhaustion, freed from being home-bound and isolated, freed from broken bones that will plague you forever, free from falls that can kill you, free to add normalcy to your life!

The trick is to recognize the point at which you begin narrowing your life and risking severe injury. That moment isn't as clear-cut as it sounds because we all tend to not want to recognize what is going on. Here is my list of subtle hints that it is time for a wheelchair:

- ✓ When you limit your fluid intake because walking to the bathroom is so tiring and risky.
- ✓ When you leave without going into a store because there is no parking space near the entrance.
- ✓ When you pay outrageous prices for groceries at the Quick Mart because a trip through a real grocery store is so tiring you can barely make it back to your car.
- ✓ When you have to use the curb cut-outs anyway because a curb is a risky climb.
- ✓ When you find your self making decisions on where to eat based on whether they have a drive-thru.
- ✓ When a trip to the refrigerator or bathroom cannot be navigated during a commercial break.
- ✓ When you regularly hear yourself saying "I'll just wait in the car".
- ✓ When you stay home because you know your family will have more fun if you are not along to slow them down or limit where they can go.
- ✓ When you sit through an entire half hour of "Saved By the Bell" because the TV remote is across the room.
- ✓ The obvious: When you fall down even while wearing the ankle-foot orthosis (brace) made to keep you from tripping. If you fall while using a walker, it is a Red Alert to get a chair NOW. It will take months to get a power chair so if you want a power chair, get started on the ordering process yesterday!

People resist going to wheelchair use for a variety of reasons; it is embarrassing to be seen in one, it is giving in to ALS, their legs will lose whatever strength they have left. News Flash: You won't look half as pathetic in a wheelchair as you will sprawled on the floor after a fall, so get over it, suck it up, etc. Using a wheelchair gives you the mobility ALS is trying to rob you of so it is definitely not giving in, it is fighting back. Your legs may lose some strength, but they will lose it anyway when (that is when, not if) you are laid up after a fall. Broken bones, concussions, sprains and torn ligaments on top of ALS weakness are very disabling, and ALS people rarely get back to where they were before the injury.



## *Manual Chairs*

Your first wheel chair will probably be a manual chair. A manual chair is convenient to take along on any outing whether you end up using it or not. Although insurance will pay for a new chair, used chairs are easily found at garage sales or Craig's List. There are two things to check before buying used: Make certain the brakes work and the tires are not worn and loose on the rims. If you find a standard manual chair hurts your butt, cut a piece of 1/2 inch plywood to fit in the seat and top that with a high-quality pressure relief cushion such as a ROHO air cushion or a gel cushion.

## *Scooters*

What about a Scooter? A scooter is a temporary solution but it gives more independence than a manual chair that you most likely can't use by yourself. It is nice to have if you can afford one. The length of time you can use a scooter limited by the need to lift your arms to reach the driving controls on the tiller, and the lack of good side support when trunk muscles weaken. Considering the short time it will work for you, a good used scooter, often a barely used one, is much less expensive. A scooter salesman will gladly encourage you to buy, telling you that he will file for Medicare/insurance and the scooter will cost you little or nothing. That is true, but doesn't take into consideration that you will graduate to needing a power chair. Medicare/insurance will look twice at a claim for a power chair if you filed for a scooter in the last couple of years. They probably will pay for it but the claim process may be drawn out with denials and more paperwork.

## *Power Wheel Chairs*

So we arrive at the pretty much inevitable power wheel chair. Many people use a recliner for comfortable seating during the day but a correctly fitted and equipped power chair is just as comfortable, provides better postural support, protection from pressure sores, helps to minimize foot and leg swelling, and is much more versatile.

Used power chairs are fairly easy to find and even newer, barely used chairs with all the necessary features are often about a tenth the price of a new one. Why? Because Medicare/insurance pay about 80% of the cost of a power chair. Properly out fitted for an ALS user, power chair costs an average of \$27,000 so it costs the user about \$2,100. When the user dies the family doesn't have to get a lot for the chair to cover what was spent on it. Putting a high price on even a top of the line, barely used chair means it is unlikely to sell.

So why not buy a good used chair? If you are not eligible for Medicare or Medicaid nor have other insurance, a used chair can be a good, affordable option, but you need to buy carefully. You must be able to sit in the chair before you buy. Ideally, before you buy a used chair, you would have an occupational therapist or wheelchair seating specialist evaluate the chair with respect to whether it is the right size for you or can be adjusted to fit. This isn't just a chair you can get out of when it gets uncomfortable. You will probably be in for most of the day. If it is too small, too big, made for someone long-waisted or short-legged, it may be adjustable to fit you but don't buy it until it is check out by a wheelchair seating company. They will also be able to tell what add-ons are available down the line such as torso supports or alternate driving controls for the brand and model. If you have Medicare/insurance they will not pay anything toward a used chair. And if you are insured, your copay on a new chair will generally be even less than the cost of a good used chair.

Aside from cost, a new chair will be built specifically to fit you, built with all the features you need, and built with adaptability for your future needs. A used chair may need adjustments and parts to fit you and that can get very expensive, especially if it wasn't a close fit to start with. You may be able to get a used power chair from the MDA Loan Closet. Type of chair and size is limited and it may take quite a while to get a suitable chair but it will be free and yours to use until you no longer need it.

The first step in buying a new chair is to find out what DME (Durable Medical Equipment) providers are covered by your Medicare/insurance policy, especially if your policy requires you to go to doctors and other providers in their network. There are not that many providers in most areas and you will want to have one within reasonable driving distance if possible. It is frustrating to have to spend hours on the road just to have an adjustment made to your chair. Some providers are good about sending someone to your house but others have such big areas to cover they can only do that if your chair problem or your condition makes it too hard for you to go to them.

It cannot be emphasized enough: Don't buy a power chair on your own. Buying without qualified help can result in a chair that doesn't fit your body and is uncomfortable, not adjustable enough to make it work for you, and not adaptable for future needs.

It is important to go through a physical therapist familiar with ALS. The therapist will know the paperwork, physicians orders, and the insurance justification needed to get all the parts and add-ons that will make the chair adaptable for you well down the road. Hopefully the therapist will work with a certified wheelchair specialist, not just a salesman. That specialist is the one who will order, assemble the chair, and do any tweaking or changing out of equipment to make it the best chair for you.

Begin by calling the local MDA office to get a clinic appointment to be evaluated for a power chair. The cost of a clinic visit is billed to your insurance and any amount not paid (or if you don't have insurance) will be absorbed by the MDA. At the clinic visit, you will see a therapist who will take you through the process of getting your chair. The therapist and usually a wheelchair specialist will evaluate not only your current strength but also your future needs based on typical ALS progression. You will be measured to get the right back rest and seat size, need for additional options such as tilt and recline, head rest, etc. will be considered, and a recommendation for the type and brand of chair made.

## **Important Power Chair Features and Considerations**

### *Tilt, Recline, Leg Lifts*

Tilt and recline and power leg lifts are features you will need. Tilt angles the chair backward for periods of pressure relief and also makes it much easier to get you scooted back in the chair so you are seated comfortably. Recline lowers the back rest and when used with the leg lifts can let you lie back in your chair with your feet up. Recline won't get you flat but when used with tilt it may. Standard power leg lifts are fine for adjusting your legs while sitting up, but when you lie back in your chair and raise the footrests, the footrests are suddenly too short! Your knees have to bend or you need a big pillow to get your heels above the footrests. Very inconvenient and hard to get comfortable! The solution is to order "articulating" leg rests. These lengthen as they lift so that your legs aren't scrunched even with the legs all the way up. Comfortable for elevating your feet to reduce swelling or just catching a nap! All this is necessary for comfort, preventing pressure sores, naps, dentist visits, and any woozy spells if you are prone to fainting.

### *Seat Cushion*

A foam cushion just isn't good enough for a full-time wheelchair user and neither is the seating that is standard on some chairs. Any seat cushion on your chair needs to be removable so that the a high-quality cushion sits on the flat metal base of the seat. The best options are a ROHO air cushion or a gel cushion. Most people prefer the ROHO. It is expensive, about \$360 on the SpinLife website, and is covered by insurance and Medicare. The people where you got the chair should be able to help you with the paperwork. You will want the 4-inch thick High Profile Cushion. It comes in many sizes to fit your chair and you need the largest size that fits in your chair. The ROHO comes with a hand pump and patch kit. New cushions have Smart Check for determining the proper inflation for a specific user. It is essentially a glorified tire pressure gauge but it something users have wanted for years! It saves a lot of the trial and error adjustments of older cushions. I find it odd that with it they are now recommending daily checks of your pressure setting. Odd because once I find the right inflation amount with my old cushion, I don't need to adjust it unless I have had to patch a leak (an extremely rare event).

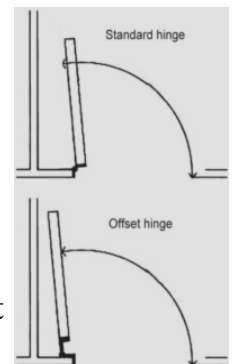
For anyone without the Smart Check, here is how to set up a ROHO air cushion. I suggest pumping the cushion up fairly full, then leaving the valve open for about 15 minutes. It won't deflate completely, just remove excess air. Then get into your chair and sit on it. It will still feel rock hard and need to be deflated more! You are not supposed to sit *ON* the cushion, but rather to sink down and sit *IN* it. Have a helper put their hand, palm down, under the bones of your butt to help check the inflation. They should be able to wiggle their fingers just a little without feeling them bottom out on the metal of the seat pan. You will have to open the valve while you are sitting on the ROHO to force more air out -- and you will probably be surprised at how much air has to be pushed out before the cushion is comfortable. If you find you have let out too much air and are bottoming out, just use the pump to add more. That can be done even while you are sitting on the cushion. It will probably take a couple of tries to get it inflated/deflated to where it is comfortable, but once you do it won't need adjusting for months. All this fussing around to get the pressure right is necessary and well worth the effort!

### *Attendant Controls*

Attendant controls allow someone else to drive the chair and are very helpful. Attendant controls are usually mounted on the back of the chair. That means an attendant cannot hold a door open while driving you through. Backing through does help but the door is going to be closing before you are all the way through. Simple solution: Buy a rubber door stop and keep it on your chair.

### *Total Chair Width*

The width of the chair is so important when it comes to getting through doorways. The chair itself may not be a problem, but contoured arm rests, padded elbow stops, leg rests and many other add-ons can add inches to the width. The width of the chair is determined by your size, but the choice of the brand and how accessories do make a difference. Measure your door frames and determine what if any modifications can be done to narrow doors. (Off set hinges add about 2 inches of door frame space!) The space at the approach to the door is as important as the width of the door. If you don't hit the threshold dead straight on, the chair will turn in the doorway .



### *Head Rest*

At some point, you will need a head rest. Most chairs accept any type or brand of head rest so this isn't a concern in selecting a chair.

### *Joystick*

Most of us start with a standard joystick but may need something different later on. The electronics for your chair should accept other driving control systems.

### *Tires*

Solid tires are rubber and/foam filled. They are the least expensive, long lasting, cannot go flat, and maintenance free. They do not cushion bumps so give a rougher ride outdoors but allow the chair to roll and turn easily. Pneumatic tires are air filled so absorb bumps better for a softer ride. They can be punctured and require regular maintenance. Semi solid tires are air and foam tires with solid inserts to prevent punctures. They provide medium cushioning and require some maintenance and replacement. Most of us will do fine on solid tires as long as we can avoid cobble stones!

### *Space for Bi-level or Invasive Ventilator*

Consider is how a Bi-level Ventilator or vent can be mounted on the chair in the future. If the chair already has the mechanics for reclining taking up space on the back of the seat, adding a Bi-level or Invasive Ventilator tray or holder can make the chair considerably longer and less able to maneuver in tight spots. You will be assured that the equipment can be mounted but insist on seeing how it is mounted and what it adds to the length. It is a nasty shock to have selected your wheelchair and purchased a vehicle that the wheelchair fits in easily, only to find out when you need breathing equipment that the getting the chair in is a major effort.

### *Elevated Seat Lift*

A power lift seat will not be covered by Medicare and costs about \$2,000. It is terrific for helping you stand to transfer independently from your chair, and when the time comes when you need to be lifted to stand it is a back saver for your caregivers. When ALS is progressing rapidly, the money spent on this feature may be better spent on an overhead lift system.

### *Standing Chair*

Medicare/insurance is very unlikely to pay for it. It is nice to be able to rise up to have more face to face conversation with people who are standing, but the other two reasons to have the elevated seat are of short term use in ALS. The standing option makes standing transfers easier for patients and caregivers, but standing transfers won't work later on. The other reason for an elevating seat is to be able to reach things on higher shelves or work at the kitchen counter top. If you love to cook and still have strong arms, it may be worth the cost to you but when arm weakness develops, you won't be reaching for stuff anyway. Being able to stand for the purpose of weight bearing would be great for the medical purpose of slowing the development of osteoporosis. A standing chair would also make it possible to attend concerts and events where everyone stands up in front of you. ("Bruce! Bruce!") I don't think insurance will see that as a medical necessity though!

### *Rear, mid, or front wheel drive*

The big decision with a power chair is the type of drive; rear, mid, or front wheel drive. There are pros and cons to each type of drive. Ideally, you would have the opportunity to try each type in your home and outdoors on the terrain you are likely to encounter. That is a joke. Few vendors have a demo chair of each type for you to try. Most will be more likely to

have one or no demo chairs. There are some differences between drives that can help determine what should work best for you.

- ◆ A rear wheel drive is very good for outdoor use off sidewalks. The push provided by the rear wheel drive can get it up and over most smaller obstacles allowing it to travel well over grass, snow, rough ground and trails as well as up driveway curbs. It handles higher speed smoothly, making it the best choice if you want a chair that will travel any distance efficiently and quickly. Indoors, a rear wheel drive has a somewhat larger turning radius but works well. It moves smoothly without the lurching that is synonymous with mid-wheel drive. It steers intuitively making it easy for any caregiver or friend to drive using an attendant control joystick mounted on the back of the seat.
- ◆ Front wheel drive is not as common. It is the best for climbing over obstacles as high as two inches or more such as curbs and does so without taking the bump at high speed. It can handle snow, gravel, and rough terrain but tends to fish tail at higher speeds so has lower speed built in.
- ◆ The drive most often recommended for those whose main use will be indoors is the mid-wheel drive. It has the smallest turning radius so it can turn in an area slightly larger than the chair itself. Outdoors a mid-wheel drive chair can get hung up on uneven, soft, or snow covered ground or on badly cracked sidewalks or streets. If the front or rear casters are on top of a high spot it can leave the chair resting on the other set of casters with the drive wheels off the ground and spinning uselessly. Climbing any curb cut out that is not fairly level and gradual can cause this and require a ramp. Driveway curbs fit this category but unlike sidewalk curb cuts, are wide enough to be climbed by driving up at an angle. Advancements in mid-wheel caster design are resolving these problems very impressively.

A problem with mid-wheel drive that never seems to be mentioned by the manufacturers but does come up in discussions by mid-wheel drive owners is called Caster Jerk. This is not the typical flutter of any caster. It is a jerk or lurch to the side.

Any wheel chair has some caster jerking as the casters swivel 180 degrees from forward to backward. There is resistance to the swivel until it reaches 90 degrees and then it finishes the swivel quickly causing a little jerk in direction. The movement is slight and soon ignored.

With a mid-wheel drive, however, there are four casters attempting to change direction 180 degrees. The jerk is accentuated and can be a problematic lurch to the side. This is minor and easily accommodated to in average size rooms where the casters have enough distance to travel to move more smoothly through the swivel. This can be adjusted with steering control settings or may require moving the entire seat on the base. It cannot be eliminated entirely.

In small spaces, it becomes a significant problem if it is necessary to back up, turn, and pull forward again to position the chair correctly. When there isn't enough space/distance for the casters to swivel smoothly, they jerk the chair quite powerfully to one side. The jerk occurs even after the joystick is released and even if you attempt to steer to the opposite side. Perhaps the most frustrating aspect of caster jerk is that once you are in a tight space you can't adjust the position of the chair without repeated jerks. Times when you are likely to encounter jerking include maneuvering in a small bathroom to position the chair accurately beside the toilet or at the sink, at a computer desk where you need to be centered and straight on, in a van where you need to face

forward in alignment with the tie downs, in doctors and dentists exam rooms, restaurants and buildings with small entries.

The jerking problem is one that most users become accustomed to, and they love their mid-wheel chairs, but will admit jerking does occur and is annoying. People for whom the mid-wheel drive is their first chair accept the problem as part of wheel chair life. Switching from rear wheel to mid-wheel is more difficult. The jerking is more obvious and can make rooms and spaces designed for a rear wheel drive very difficult in a mid-wheel drive.

For people who can drive themselves, a mid-wheel works well, but when they deteriorate and an attendant must drive, it can be problematic. Each caregiver must work with the chair often enough to overcome the learning curve for basic driving and, depending on the home layout, for tight spaces.

### *Selecting a Vendor*

You probably won't have too many choices about which vendor (durable medical equipment provider) you get the chair through, but consider the distance you have to travel to have it worked on. Having to travel hours just to have a speed setting changed is annoying. Not all vendors service all brands.

### *Paperwork*

Once all this is decided the paperwork begins.

1. The therapist will complete the forms to justify your need for a power chair and all the add-ons (tilt, recline, etc.) Knowing the right codes and buzz words to use is critical in the process of getting your chair. Errors or omissions will delay insurance approval by a couple of months.
2. Next the paperwork must be signed off on by the durable medical equipment provider (vendor) who will actually order the chair from the manufacturer.
3. A physician signs the papers.
4. The papers go back to the therapist or vendor to be sent to Medicare/insurance company.

Because of this process, the traveling paperwork can get stalled anywhere along the way. Be a pest and call the therapist in a couple of weeks to find out how far the paperwork has gotten and continue to check on it until it has been sent to the insurance company. The chair won't be ordered from the manufacturer until Medicare/insurance approval is received by the vendor, so once you are told that the paperwork has gone to Medicare/insurance, call the equipment provider every month to track progress. Expect a minimum of three months and more likely at least six months to get the chair approved, built, delivered, and final adjustments made.

Hopefully you will be able to get a van to transport you in your wheelchair. Ideally, you will be able to try the two out together but that isn't usually the case. If you are tall or require an extra wide chair, standard van interior space and headroom may not match the chair's requirements, so don't rush to buy a van until you have the chair to try in it.

The chair will need to be fastened down inside the van. One thing to consider is where the tie downs attach to the chair. Some chairs have the tie down spots up on the seating section of the chair. This might have some safety advantage in an accident but it does require re-doing tie downs if you want to tilt or recline the chair while out and about. Tie downs on the base of the chair don't affect seating position changes.

When your chair arrives expect to spend several hours with the specialist getting it set up, fitted to you, and controller speeds adjusted. Bring along someone who will be your designated mechanic for any adjustments you don't want to come back into the shop for -- like fixing a skewed foot rest after plowing into a wall. (A dry wall handyman is nice to have too!)

Never, ever sign anything to say you accept the chair until you have taken it home and tested it there in every room where you can anticipate ever using it. Test it on your wheelchair ramp, on grass, gravel, and slopes. Drive it into a van and make certain you can turn to face the front. Test it in restaurant entries and dentist and doctors rooms. Remember that there will be a day when you won't be able to get out of that chair to walk, hobble, or be hauled around an obstacle. Only sign for it when you are satisfied that it can get you where you want to go.

### *Choosing a Van*

#### *Minivan, SUV, or full size Van?*

##### 1. Minivans

- More appealing to those who don't want to drive a "truck".
- Will fit in garages and parking ramps.
- Uses a side or rear entry ramp rather than a lift.
- Require lowering of the floor for the ramp.
- Lowered floors can bottom out on rough roads or driveways.
- In a side entry minivan the entire floor is lowered.
- In a rear entry minivan only the floor between the rear wheels and up to the front seats is lowered leaving a channel for the wheelchair.
- Rear entry limits the seating available for other passengers.
- Less interior space and less headroom makes it more difficult to use with a power chair, especially for a tall person.

##### 2. SUVs

- Some power chairs may not have the turning radius needed to get turned facing forward in SUVs. Most newer power chairs pivot closer to their center however, and should work but testing before buying is important.
- A side entry SUV may have only the section of the floor between the front and rear seat lowered. This leaves little room for turning a power chair to face the front and rear seat passengers with feet dangling above the lowered floor.
- In a rear entry SUV only the floor between the rear wheels and up to the front seats is lowered leaving a channel for the wheelchair.
- Rear entry limits the seating available for other passengers.
- Less interior space and less headroom makes it more difficult to use with a power chair, especially for a tall person.

##### 2. Full sized vans

- More space for wheelchair and other gear.
- May not fit in some garages or parking ramps. Raised roof version will not fit.
- Uses a side or rear entry lift.
- Must have floor lowered and/or roof raised to accommodate wheelchair and lift.
- Consider size of engine cowling between front seats. Can make it very difficult to move back to the passenger area if wheelchair passenger needs help.
- Allows seating for passengers.

- Available in longer lengths for even more seating or storage.
3. Other options:
- Dodge Sprinter: Available from manufacturer in two roof heights, 64 or 72 inches of headroom, no roof raising/floor lowering conversion expense, just add a lift. High door height for even the tallest wheelchair user. Three lengths available. Ideal for traveling. Excellent visibility for wheelchair passenger. Side or rear entry.
  - Ford Transit Connect. A small size utility van that is becoming popular for wheelchair use. Rear entry only. Fold down ramp rather than lift. Must have floor lowered. Two lengths available. Shorter length has seating for driver and one or two passengers. Longer length allows more passenger seating but puts the wheelchair in the 3rd row, and space may not be long enough for a power chair.

### ***Lower the floor or raise the roof?***

- Sitting in a wheelchair puts the person sitting too high to see much out the window beyond the edge of the road. Raising the roof doesn't add height to the windows. Lowering the floor is somewhat better but the wheelchair passengers view is still quite limited.
- In order for a caregiver to stand up in the van, a real help but not essential, both floor lowering and roof raising must be done. A lowered floor or raised roof are generally not available as factory options and must be done by a van conversion shop.

### ***Side or rear wheelchair entry?***

1. Rear entry
  - Allows the wheelchair user to get in without maneuvering to turn the chair.
  - Rear entry limits the seating available for other passengers.
  - Rear entry eliminates the problem of being blocked from using the lift by other parked cars, but requires loading and unloading in traffic lane of a parking lot.
2. Side entry
  - Requires turning the chair to face the front. Riding sideways is unsafe as well as nauseating.
  - Requires about 8 feet of space, and another car parking too close can require moving the van to get back in. That is a mere nuisance if you are a wheelchair passenger, but if you are still driving yourself it leaves you stranded.

### ***Lift or ramp?***

*Minivans use ramps.*

- Ramps can be mounted inside the van or positioned under the floor.
- Manual or automatic.
- Takes up some space in passenger area and may partially cover the window although some (\$\$) fold horizontally or vertically for better visibility.
- May prevent front passenger seat from being able to move back (reducing leg room) or reclining.

*Full size vans and pickups use lifts.*

- Lifts are automatic (powered) and are wired into the vehicle's electrical system and can be operated even when the vehicle is not running.
- Can be operated manually if controller fails.
- Lifts can be mounted inside the van or positioned under the floor.
- Semiautomatic raises/lowers the lift with a switch located on the lift, requires a caregiver to open the door.



- Fully Automatic opens door, raises/lowers the lift, and closes the door with switches located on the lift. Can be used independently if hand/arm strength allows.
- Hand-held remote control can be used instead of switches. Requires full attention and caution to be used safely.

1. Folding

- Takes up some space in passenger area and may partially cover the window although some (\$\$) fold horizontally or vertically for better visibility.
- May prevent front passenger seat from being able to move back (reducing leg room) or reclining.

2. Under the Floor Slide Out

- Doesn't take up passenger space or block the window but may take 1.5 inches away from the headroom, a small but critical amount.
- Harder to deploy manually if something happens to the controller.
- May require modifying exhaust system, gas tank.
- Enclosed but somewhat more exposed to water, snow, salt.

## ***Low Cost Ways to Keep Computing***

Being able to use a computer has been such a positive aspect of my life with ALS that I want to share the tricks and tools I use to keep computing.

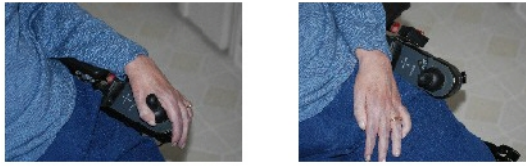
### *Step One: The Right Computer Desk*

The first thought in computer access for the disabled is usually about finding hardware and software, but accessibility has to begin with even more basic problem solving; finding a desk that will allow you to use your computer!

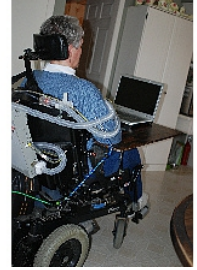
Over the years I have been through any number of rigged up computer setups as I fought to keep ahead of my ever-increasing weakness. Every setup turned out to be temporary -- and looked it! I finally had enough of crappy looking desks and set out to find a workable, adaptable, computer work center that looked like furniture. The desk I came up with is made from Sauder furniture and is the one I should have started out with because it is adaptable through many stages of disability. I was already in a power wheel chair when I came up with the idea for the desk so it was built to wheelchair height, but could be made at standard desk height and elevated later using leftover, matching wood to maintain its good looks.



The first requirement of a desk is support for weak arms. It amazes me that very, very few computer desks for wheelchair users have this feature! My arm support is cheap, easy to make, uncomplicated, and roomy. I use a piece of plywood with a deep cutout so that it wraps around my waist and extends far enough back to support my elbows. This tray slides out from under the desktop and rests on my wheelchair arm rests, providing support at just the right height for shoulder comfort. At the time I designed this desk, I was still able to reach the tray and pull it out, as well as use the keyboard. I can't do either of those things now, but the pull out tray is easy for my caregivers. (I have to rotate the joystick for my wheelchair a bit to fit under the tray.)



I also have a portable tray made exactly like my desk tray. It is held onto my chair with two short bungee cords (the blue straps) that hook through two small holes drilled into the back edge of the tray and then down to hook on the wheelchair frame. It is a really inexpensive, quick to set up, roomy solution for computing anywhere I go, but requires assistance to set up even if you have arm strength.



Arm supports recommended by Occupational Therapists are from medical/rehabilitation suppliers and are very expensive. They attach to the wheelchair which can be a real nuisance for transfers. The one advantage of this type is that they are available with springs or large rubber bands to help you raise your arms to feed yourself. This feature isn't needed for computer access though.



Commercial arm support devices are available with the most affordable types found on web sites selling ergonomic devices for computer users. Besides being inexpensive, they are generally made to clamp onto the desk rather than the chair. Some combine the arm support with an attached mouse pad support.



Perhaps the most affordable desk to use with arm supports is an adjustable height drafting table. These can be found for as low as \$110. In addition to being inexpensive, they are small enough for cramped quarters, require minimal assembly, and adapt to any wheelchair height. The downside is that they have no space for a printer and other peripherals.



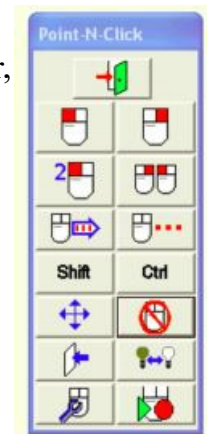
### *Step Two: Tame the Mouse*

Before making the switch from mouse to some type of eye gaze cursor controller, there are some things that can be done to prolong your mousing ability:

1. Reduce the need to click the mouse.



- Point-N-Click (freeware) can be set to automatically left click whenever you place your cursor over a clickable item. That alone saves a gazillion clicks but isn't the only option.
- You can quickly (and without clicking) switch back and forth to right click, double click, click and drag, select multiple items from a list, and more.



- It doesn't clutter your view of whatever is up on your monitor. It hides off to the side of the screen and the control buttons are made visible by putting the cursor on the little edge that remains on the screen.
- You can have it display only a few click options or many.
- Excellent for scrolling because you can adjust scrolling speed as needed.
- It works with a mouse or eye gaze equipment.

## 2. Change your mouse pad.

I use an over-sized pad so that I don't can't push the mouse over the edge and have to struggle to get it back on the pad. XTracPads Ripper XL™ v2 is fantastic! It is 14 x 17 3/4 wide and has a friction free smooth cloth surface that maximizes the strength I have left.

## 3. Free up your elbows.

For quite a while I used a furniture moving pad such as a Moving Men pad to allow smoother elbow turning, pushing, and pulling the mouse. More recently I came up with an elbow roller that works even better. Get 4 small rolling casters from Banana Robotics. Cut about a 4X6 piece of thin wood or non-flexing plastic. (It could be smaller but I found that the small size cut off circulation and made my arm go to sleep.) Attach the rollers with small screws or glue. Add a pad of cushioning foam and your mouse will skate easily!



## 4. Dress appropriately for your office.

- Avoid bulky clothing and rough fabrics that limit arm movement.
- Get the right mouse and stick to it.

I use a simple optical mouse and prefer one without a scroll button which just gets in the way, but they are hard to find. A wireless mouse is just too heavy to push around. I have had to switch to my left hand, and need help to lift my fingers up onto the mouse. When the humidity is low or my hands are cold and stiff, a piece of tape helps hold my fingers on the mouse. Velcro tabs on the right and left buttons keep my fingers in place.

## 5. Nail that tail.

I have found that the mouse cord hanging down will quickly pull the mouse sideways so the cord is tucked under the keyboard or taped to the desktop to hold it in place.

### *Step Three: Ditch the Keyboard*

You don't need to be able to use a keyboard to use your computer, and you don't need to buy any software! The software programs you need are **free**. Before you assume that purchased programs would be better, consider the fact that this website is done using these freebies. I also do photo editing and have a website for that, Free Photo Fix Magic.

So what are these great programs?

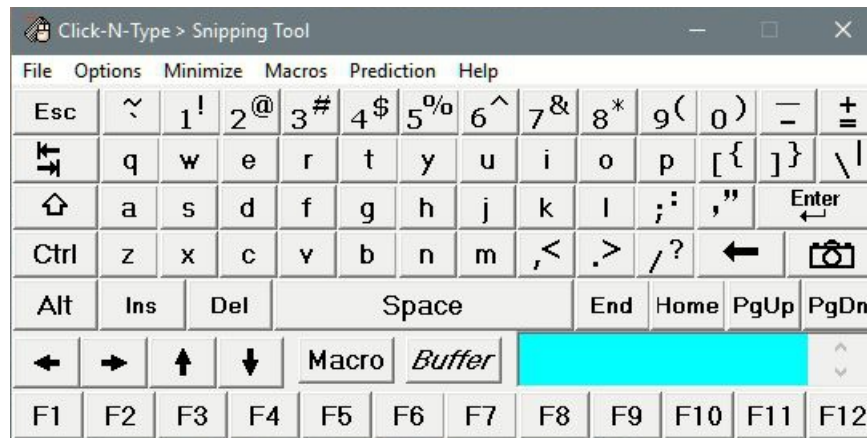
#### **Click-N-Type**

The first program I rely on is a free onscreen keyboard called Click-N-Type. It puts a keyboard on the screen and as you move the cursor over the keys, it types for you -- or lets you control your computer with any keyboard command (Control/Shift, F4, etc.). You never need to struggle with your keyboard again!

Several great features make this freebie indispensable:

- You don't have to click the mouse for each letter. Just set it on auto click and it will click as you pause the cursor over a key.

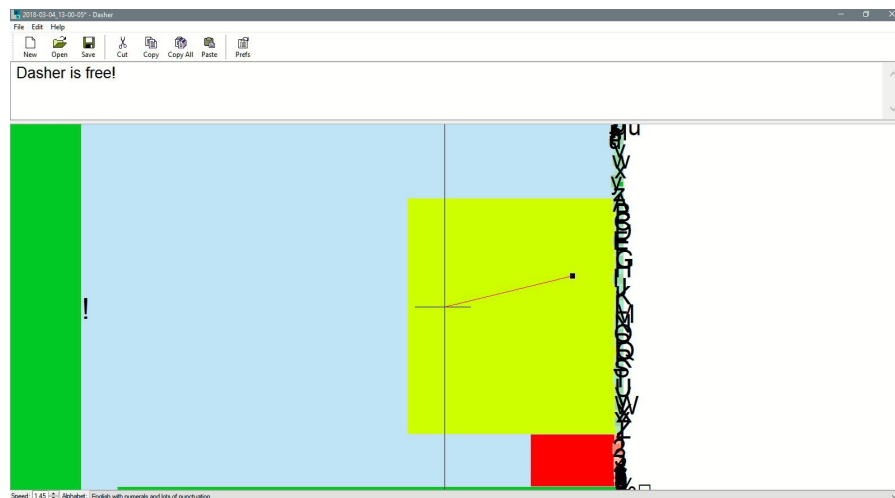
- You can set it with Macros that type frequently used words, phrases, or sentences with just two clicks.
- It works with a mouse or eye gaze equipment.



- It offers several keyboard layouts; standard Qwerty, alphabetical and more. You can also to design your own. I have created custom layouts to speed up typing, code html, or use just the number keys. The one shown here groups the alphabet in the center rather than in two long lines, making it faster to find the desired letter. The most often used letters are centered, making it even faster. Having a couple of the blank spaces lets you pause in your typing without leaving the program.
- It has Word Prediction to speed up typing.

## 2. Dasher

- When I want to compose an email or a bit of literary genius, I Use Dasher, another freeware program. It allows much faster typing because it isn't hunt and peck.



- Dasher is like playing a race car game -- you just drive the cursor, pointing it down the road to the next letter you want to type.
- With just a little practice you can move at speeds nearing that of ordinary typing. The speed allows you to work at composing, not just plunking out letters!
- Dasher has the option to speak what you type!
- It works with a mouse or eye gaze equipment.

### *Step Four: Move the Monitor*

Even the slightest amount of neck weakness can make using the computer an effort. Neck muscles tire quickly if not supported, but leaning back to use a head rest changes the angle of view, causing eye strain. I thought I needed new glasses when I couldn't read anything but the largest type. Then I discovered that all I need was a booster seat for the monitor! By raising my desktop monitor up about 5 inches, I can rest my head and still see the screen at the right angle. I expect that at some point I will have to find a way to compensate for a more reclined position by both raising the monitor and tilting it forward. But for now, my monitor is on a sturdy riser. My laptop required a much higher boost to bring it up to a comfortable eye level and a collapsible, completely adjustable for both height and angle monitor stand has been terrific. Easy to adjust, more and taller heights possible, stable without clamps or bolts, and the laptop doesn't need to be attached to it. Several brands are available.





## *ALS Information for Caregivers*

I was asked to do an educational program for a local nursing home on the care of the ALS patient. My lung capacity had gotten too low to allow me to speak loudly and long enough to do this in person, so I offered to make it a written in-service. The focus is on the care issues presented by ALS patients that nursing home staff may be less familiar with so the in-service does not cover some aspects of care common in nursing homes, such as tube feedings.

Corrections or suggestions for improvement would be much appreciated, especially comments from PALs and CALs who are familiar with the educational needs of nursing home or home caregivers regarding ALS.

If you feel that the information would be of help to any caregivers, feel free to copy it and share it.

### *To the Staff Instructor*

Feel free to use this information in any way you feel it will be helpful. Add, delete, rearrange as needed and pass it on to anyone else who might find it useful. I started to add goals and objectives but decided my nursing educator ways are probably getting a bit out-dated. I will leave it to you to add the necessary spit and polish needed to make this a "real" in-service! Please let me know if there are any clarifications needed, more specific aspects of ALS you would like covered or if there is anything else I can do to help.

### *To the Staff*

I apologize in advance to readers who may find this information rather dry and boring. If I were able to deliver it in person, it would be much less 'textbook' and a lot more interesting! Unfortunately, ALS has made speaking to groups difficult for me so you will just have to wade through this without benefit of my charismatic speaking style, sparkling wit, warmth and bubbly personality! (And doughnuts. I always brought doughnuts!)

To answer any questions on my credentials . . . I am a retired RN. I worked general Med-Surg units, then 5 years in Critical Care where I began specializing in Neuro and earned Neuro Certification (CNRN). I was diagnosed with ALS in 1985 at the age of 37. When my hospital opened a Neuro Unit, I was selected for the position of Neuroscience Educator. In that position, I was able to continue working even after I began using a wheelchair. By 1995 arm weakness and fatigue made it too difficult to continue and I retired.

I have been incredibly fortunate in that my ALS has progressed very, very slowly, and, unlike most ALS patients, I have been able to enjoy the years since my retirement. I spend my days at my desk with computer, TV, stereo, and 3 cats to keep me entertained, and a wonderful husband to care for me. My computer has kept me occupied, entertained and even productive. I play games and create photo album CD's, screen savers, brochures, newsletters, etc., for family and friends.

Through the Internet, I communicate daily with other ALS patients, keep up on research and treatments and care issues. I have my own ALS web site *ALS From Both Sides* which focuses on nursing care, and on practical solutions for the daily problems of ALS.

If you have any questions about ALS or care of ALS patients, I will be glad to answer if I can, or look for the answers for you.

Diane Huberty

dianehuberty@frontier.com

## *ALS: The Disease Process*

### *What is ALS?*

ALS is the abbreviation for "Amyotrophic Lateral Sclerosis." "A-myo-trophic" comes from the Greek language. "A" means no or negative. "Myo" refers to muscle, and "Trophic" means nutrition or stimulation. When a muscle has no stimulation, it "atrophies" or wastes away. "Lateral" identifies the area of the spinal cord where the pathways for motor nerves, those that innervate the muscles, are located. As this area degenerates it leads to scarring or hardening, "sclerosis," in the region.

ALS is a progressive neuromuscular disease. It attacks motor neurons in the brain and spinal cord which transmit signals to the voluntary muscles throughout the body. When motor neurons die as a result of ALS, the ability of the brain to control muscle movement is lost. Weakness spreads to all the voluntary muscles and progresses to paralysis. ALS does not affect sensory nerves so there is no loss of feeling in the paralyzed areas. ALS only affect motor pathways in the brain so awareness, thought and intelligence are not generally affected even after paralysis makes it impossible for the patient to communicate.

In the United States, ALS is also known as Lou Gehrig's Disease after the baseball player who died from ALS. In other countries it is often called "Motor Neuron Disease." This leads to some confusion because there are several other forms of motor neuron diseases such as SMA (Spinal Muscular Atrophy) and Kennedy's Disease. ALS is actually just one of the motor neuron diseases. Currently ALS is categorized in two forms, Familial and Sporadic. About 10% of ALS patients have Familial ALS which is an inherited disease. The majority have Sporadic ALS in which no family history can be found. There is no difference in the symptoms and progression of the familial and the sporadic forms of the illness.

### *Cause*

Even though changes in the levels of a substance called glutamate have been identified as being at least a part of the process that kills the motor neurons, the cause of the increased glutamate is not fully understood. Research continues to look for links between ALS, genetics, toxins, antioxidants, viruses, and autoimmunity. Although some scientists believe it is possible that ALS is caused by a slow-acting or latent virus, no such virus has ever been identified, and there is no increased incidence among family members, care givers or medical personnel because they are in close contact with ALS patients. Many researchers believe that in the end it will be found that there are many ways in which the process of motor neuron degeneration can be triggered and therefore several possible causes of ALS.

### *Who Gets ALS?*

ALS occurs throughout the world with no racial, ethnic or socioeconomic boundaries. Most people who develop ALS are between the ages of 40 and 70. It is not uncommon among people in their twenties and thirties however. Men are affected slightly more frequently than women.

ALS is not an extremely rare disease. The incidence is about 2 new cases per 100,000 every year. Because life expectancy is so short, the number of ALS patients alive at any time is low, but about 5,000 people in the U.S. are newly diagnosed with ALS each year.

## *Signs and Symptoms of ALS*

Even before weakness is noted, muscle twitches, (fasciculations) are common. Other patients notice a stiffness in their arms and legs. Many have increased muscle cramping. Some people first experience weakness in their arms or legs. This is referred to as limb-onset ALS. Arm weakness is somewhat more common and begins with a weakening of the grip and fumbling fingers. Leg weakness causes fatigue when walking, difficulty climbing stairs, stumbling. For others, the first symptoms may involve problems with speaking or swallowing: Slurred speech, a nasal tone, or choking easily. This is bulbar-onset ALS.

As the disease progresses, the weakness becomes more severe and spreads to other areas of the body. Although one side of the body may be a little ahead of the other in deteriorating, ALS is usually quite symmetrical. Eventually, arms and legs, swallowing and speech and breathing are all affected. As the muscle weaken, affected areas become thin and wasted. Muscle cramping and twitching eases as the weakness turns into paralysis, but the twitching and cramping moves on to newly affected muscles. In some patients, the stiffness noted earlier increases to severe spasticity and hyperactive reflexes.

Although the areas of the body first affected and the rate of progression varies, the progression of the disease is generally steady. Plateaus have been documented but are often more a matter of function than an actual delay in the spreading of the weakness. For example, a patient may seem to plateau after beginning to use a wheelchair. Because he is no longer attempting to walk, further weakness of his legs is less notable. Involuntary muscles are not affected, so the heart, bladder, and bowel are not directly affected.

The senses, including vision, hearing, and touch, are not affected. Although intelligence and thought are not generally affected, some ALS patients do experience a problem with their emotional responses sometimes referred to as "pseudobulbar emotionality." They find themselves bursting into tears or into helpless laughter even though they are not thinking or feeling overly sad or amused. The mechanism for this misdirected response is unclear and can be very distressing and embarrassing for the patient. It is often assumed that weeping spells are due to depression since depression is certainly common in ALS patients. However, if inappropriate laughter is also seen, this emotional lability is probably not an indication of emotional problems but rather a physical "short circuit" in the pathways between emotional thought and the motor responses.

A small percentage of ALS patients do experience a concurrent progressive dementia. The dementia is not considered part of the usual ALS disease process but rather an added complication in which other areas of the brain are atypically susceptible to the process that is destroying the motor neurons. In these patients, personality changes, mood swings, irritability, unreasonableness occur. Early on it can be very difficult to differentiate between a possible dementia and an inability to cope with the stresses of the disease. If dementia occurs, it is usually obvious before the patient loses the ability to communicate so it should never be assumed that a late stage, non-communicating patient has become demented or vegetative.

## *Diagnosis*

At present there is no definitive means of diagnosis of ALS. Most diagnoses are made by eliminating all other possibilities -- ailments whose symptoms resemble those of ALS. Neurologists use a number of clinical tests to establish a profile, including blood testing, EMG, MRI, etc.



## *Life Expectancy*

Average life expectancy for ALS patients is 2-5 years with the cause of death being respiratory failure, most often hastened by pneumonia. Weakening of the inspiratory and expiratory muscles compromises the ability to cough and clear pulmonary secretions. Respiratory failure usually results from an upper-respiratory infection that develops into pneumonia. When swallowing problems occur, aspiration greatly increases the incidence of repeated pneumonia and respiratory failure can occur long before the patient actually reaches the point where his respiratory muscles are too weak to sustain breathing.

Just as the onset and progression of extremity weakness and swallowing/speech problems varies, so does the onset of ventilatory muscle weakness. It may occur early on before arms and/or legs are paralyzed, or later. It may precede, coincide with, or follow the onset of swallowing and speech problems.

While the average life expectancy for ALS patients is 2-5, twenty percent of patients will live more than five years. Up to 10% will survive more than ten years. A very small percentage progress very slowly and may survive as long as 15 or 20 years or more.

It is important to note that these figures are based on patients who do not go on Bi-level Ventilator or a full ventilator. Patients who opt for tube feedings and a ventilator when swallowing and respiratory muscles fail can generally be maintained for many more years. Life expectancy among these patients has not been reported, but the patient's age, other health problems and the quality of nursing care available will affect life expectancy.

A decision to go on a ventilator does not halt the disease progress, however. Paralysis will continue to progress and all movement including facial expression, eye movement, blinking may eventually be lost. The patient can become "Locked In", meaning that a fully alert and aware mind is trapped in a body that won't allow communication with care givers.

## *Care of the ALS Patient*

ALS is a disease that has no cure and no medical treatment beyond supportive care. Therefore, the focus is on providing nursing care that helps the patient and family cope with the ever increasing disability and deteriorating breathing, maximizes communication, maintains comfort, and prevents further complications of immobility. The list of potential problems and nursing interventions for any immobilized patient is long and rather than reiterate common nursing interventions for common problems, the focus here will be on those that are additional or need to be tailored to meet the specific needs of the ALS patient, particularly those that apply to patients in later stages of ALS.

## *Communication*

Although the loss of the ability to communicate is not a life threatening complication nor physically painful, it is perhaps the most emotionally and mentally devastating aspect of the disease, the most frightening, and surely the most frustrating. The loss of input into one's own care represents a loss of independence that makes paralysis seem a minor problem. The degree of helplessness is frightening. Most frustrating of all is the fact that this problem is "treatable" -- not by medicine but by technology -- yet many patients are not being helped. For example, finding a nurse call system that the patient can activate represents the very minimum in care and yet all too often it is not done.

A lack of information about the technology available, intimidation by high tech equipment, and high cost all combine to prevent ALS patients from getting the needed equipment. A simple Internet search will turn up several suppliers for the equipment. Early use of the equipment will allow the ALS patient to become comfortable and proficient with it before it is desperately needed.

In 2001 Medicare began covering the cost of some communication aids. With the help and active support of care givers, few if any ALS patients need to be left "Locked In" by the disease.

If the problem is primarily lack of breath control for speaking, equipment is available to magnify a weak whisper into audible speech. If the problem is paralysis of the muscles needed to form words, magnification won't help, but as long as the patient retains the ability to move nearly any body part -- hand, knee, foot, mouth, eye brow -- that small movement can unlock the potential for at least basic communication.

There are a multitude of communication devices available, ranging from simple nurse call buttons to portable boxes that can speak a few pre-recorded words/messages, to larger ones that allow the patient to "type" in his own messages and even have it spoken aloud. Any of these can be operated using easy to press "switches" that replace hard to press buttons. These are called "switches" for their electronic function but are actually simply modified push buttons. Some are extra sensitive to pressure and can be activated with slight pressure from a hand, foot, knee, elbow, etc. Some are extra large for easy pressing by clumsy hands that cannot handle small buttons. Others switches are activated by slight movement rather than pressure. A tilt of the head, blink, lift of an eyebrow can be used. Mouth/breath control by "sip" or "puff" is also possible.

Ordinary laptop and desktop computers can also be set up for use by patients with near total paralysis. One type of set up allows the user to move the cursor simply by moving his eyes across the monitor screen. The mouse is replaced by whatever type of switch the patient finds easiest to use. Although it is a slower process than for other computer users, with this equipment an ALS patient has the potential for communication, entertainment, and even productivity. The fact that this technology is readily available and yet so many paralyzed people are left unable to communicate their simplest and most basic needs to their care givers is a tragedy.

What can a care giver do when the equipment is not available or the patient cannot use it anymore? Assuming that every effort has been made to get the equipment or further modify it to fit the patients deteriorating strength, the care giver can only fall back on low-tech aids such as letter boards. For a patient whose only movement is eye gaze, a clear plexiglass board with minimal words or symbols arranged on it is easy to make and works best for simple and fast communication. Facing the patient with the board held up between them, the caregiver can follow the gaze of the patient as he looks look toward the word. A number of boards can be made, each dealing with specific situations -- activities, positioning, comfort, etc. A board set up with the alphabet can be used to allow the patient to spell out his own messages for specific communication.

If even this method fails, the care giver can only go about providing care while remembering that inside that motionless body and expressionless face, there is still a person who sees, hears, understands, feels. Provide distractions -- TV, books on tape, time out of his room. And above all, talk to him, not about him.

### *Pseudobulbar Emotionalism and Depression*

Pseudobulbar Emotionalism (emotional lability) is not a mood disorder but does generally respond to amitriptyline (Elavil) or fluvoxamine (Luvox). Depression is common as it is in any devastating disease and needs to be treated if persistent. Remember that the lack of facial

expression may be due to weakness of the facial muscles, not depression. Nor does lack of expression indicate anger or ingratitude! We rely so much on non-verbal response that an expressionless patient seems angry, snooty, or depressed. It is amazingly frustrating, even irritating, to care for a patient who never smiles a thank you, never reacts to your best bedside chat. All you can do is assume they are smiling on the inside!

### *Respiratory Care*

Nursing assessment of respiratory status includes all the basics (respiratory rate, depth, effort, use of accessory muscles, breath sounds, color, sputum production). One key point differs from standard assessment, however. An ALS patient with significant impairment of respiratory muscles will not have "labored respirations" as evidenced by increased depth and use of accessory muscles. (If he had the muscles for increased depth he wouldn't be in trouble!) The real indication of respiratory distress is in the rate of respirations and heart rate. An ALS patient in trouble will have rapid but shallow respirations and increased heart rate.

Although the ALS patient is at risk for respiratory crisis from pneumonia, a simple cold, and even pulmonary emboli, sudden respiratory deterioration usually follows a slow slide.

In addition to patient complaints about the room being too warm or stuffy, chest aching discomfort, difficulty breathing especially when lying down or after meals, and headaches (especially morning headaches), other indicators of deteriorating respiratory status can include lethargy, drowsiness, confusion, anxiety, irritability, loss of appetite, fatigue, depression. In short, if there is a way to feel all-around lousy, respiratory insufficiency will do it!

Although respiratory deterioration is expected and respiratory failure is generally the cause of death in ALS, there are interventions that can significantly delay the need to choose between a ventilator or death, and, more importantly, greatly improve the patient's quality of life. Unfortunately, many physicians are unfamiliar with these interventions or misinformed. For example, ALS patients are sometimes told by their doctors not to get flu shots. Flu and pneumonia vaccines are NOT known to have adverse effects on ALS patients beyond those seen in the general population, however. Although the flu vaccine will not protect the patient from all respiratory infections and the pneumonias usually seen in late stage ALS are caused by organisms not covered by the pneumonia vaccine, they will protect the patient from most common "bugs" passed around among family, friends, and care givers.

One common problem for ALS patients is the inability to cough strongly enough to clear the airway of even the normal accumulation of mucus. In addition to making certain that fluid intake is sufficient to keep the secretions thin, an over-the-counter cough medicine containing the expectorant guaifenesin can help loosen the thick phlegm. A beta blocker, such as propranolol (Inderal) or metoprolol (Toprol) may also help reduce the amount of phlegm produced.

A weak cough can be made more effective by Quad coughing (assisting a cough by applying Heimlich-like pressure as the patient coughs), giving deep breaths with an ambu-bag to improve the cough, or using a "Cough Assist" device (a device which delivers a couple of deep breaths through a mask and then abruptly reverses to negative pressure to simulate/assist a cough).

Morning headaches are often the first sign that the next step in respiratory care, a Bi-level Ventilator, commonly called BiPAP, is indicated. Even in healthy people, breathing is shallower during sleep. For people with ALS, that little extra drop in volume can mean trouble since they are breathing shallowly to start with. As the disease progresses, patients find themselves waking up

with headaches because the shallow breathing causes them to retain CO<sub>2</sub> which gives them a headache. After they wake up and begin breathing more deeply, the headache goes away.

Other patients may wake up repeatedly during the night as the shallow breathing or even apnea triggers an internal alarm that wakes the patient. This can cause a sudden awakening jolt or just restless, fitful sleep. With broken sleep, the patient is deprived of REM sleep, the stage of sleep considered the most important. Sleep deprivation causes daytime sleepiness, lethargy, anxiety, irritability, confusion, difficulty thinking clearly and remembering things as well as physical problems such as poor appetite, nausea, increased heart rate and fatigue and weakness.

At this point, use of Bi-level Ventilator non-invasive ventilation is clearly indicated. The introduction of Bi-level Ventilation in the early 1990's represented a major advance in respiratory care for neuromuscular disease giving the patient the opportunity for respiratory assistance short of a ventilator. Unlike a ventilator, no tracheostomy is needed. Bi-level Ventilation is done using a mask over the nose that can be removed when not needed. Because it does not require a trach, it does not interfere with speech or swallowing. Unlike a full ventilator however, a Bi-level Ventilator requires that the patient be able to take a breath. A Bi-level Ventilator is NOT a life support machine -- it cannot take over breathing for the patient completely. It delivers a pressurized breath of air into the lungs, then drops the pressure to allow the patient to exhale.

The most common use of this type of machine is a CPAP machine for people with sleep apnea. CPAP is not tolerated by people with ALS. They require the extra settings provided by a Bi-level Ventilator. Unfortunately, many doctors are unfamiliar with its use in neuromuscular diseases even though Bi-level Ventilation is now part of the Standard of Care/Practice Parameters for ALS patients. Too often the orders for the pressure settings are wrong, or oxygen is ordered. A Bi-level Ventilator uses only room air and that is all the ALS patient generally needs, but oxygen can be added in later stages if needed.

Pulmonary studies of FVC (Forced Vital Capacity) may be ordered to support the clinical evidence that Bi-level Ventilation is needed. Ideally, FVC readings should be done with the patient lying down because that is when the problem is most evident. There is no set FVC at which a Bi-level Ventilator is indicated, however. Overnight monitoring of O<sub>2</sub> Saturation can be done with a simple "clothes pin" monitor on the finger to detect drops in oxygenation from shallow breathing or apnea. ABG's may be ordered but are seldom necessary at this point unless the patient also has some other lung disease. If ABG's are done, it is important that they be done immediately on awakening: CO<sub>2</sub> levels will begin to return to normal once the patient is awake. A full Sleep Study to prove nighttime breathing problems is seldom needed in ALS.

Bi-level Ventilation is generally initiated for overnight use and most patients find that assisted breathing overnight also improves unassisted daytime breathing. This may be due to the rest given respiratory muscles during the night but probably owes as much to the restorative powers of a good night's sleep. Similarly, appetite, strength, stamina, and mental and emotional state improve.

As breathing deteriorates, a Bi-level Ventilator is used for rest periods during the day, often after meals when breathing is more difficult, and eventually, it is used continually. Bi-level Ventilation can improve quality of life while delaying the need for invasive ventilation by months or years.

Although most patients adapt quickly to the annoyances and are successful in using a Bi-level Ventilator, patients who already have significant bulbar weakness may have problems. Weakness of the jaw and lip muscles make it impossible to close the mouth. Many physicians automatically assume that ALS patients cannot use a Bi-level Ventilator for this reason but jaw support straps or full face masks that cover the mouth, as well as the nose, may work.

When the oropharyngeal muscles weaken to the point where the glottis can't close off the esophagus, the air is pushed into the stomach rather than the lungs and the patient awakens uncomfortable and having difficulty breathing from the abdominal distention. Adjustment of the Bi-level Ventilator pressures and sleeping position may help, but this is often a signal that a change to invasive ventilation is necessary.

The pros and cons of invasive ventilation should have been discussed with the patient well before the time when the decision has to be made. In addition to quality of life, the financial cost and burden of care are huge considerations in the decision about going on a ventilator.

Patients need to know that the decision to go on a ventilator is not irreversible. If at some point they wish to discontinue ventilation and be allowed to die, it can be done legally and with all the medication needed for a peaceful death.

Some people use Bi-level Ventilation as an intermediary step before going on a ventilator, others find that by the time a Bi-level Ventilator is no longer sufficient and they need to consider full (invasive) ventilation, their level of paralysis and quality of life is such that they do not want to prolong life with a ventilator.

When the patient has either rejected Bi-level Ventilator or has used it as long as possible but elects not to go on to a ventilator, the focus is on measures to reduce the discomfort and anxiety of insufficient respirations. For most patients, this end stage is peaceful with slowly deteriorating level of consciousness and death. For others, it can be a nightmare of slow suffocation. Medications such as Ativan are given for anxiety and opiates, generally morphine, can be added if dyspnea is severe or constant. Morphine diminishes the respiratory drive that causes "air hunger" as well as relieving the discomforts of joint pain that is usually significant by this stage. Thorazine may also be used for restlessness.

Although these medications are used freely without (pointless) concern about addiction, the goal is not to stop respirations but only to ease discomfort. The use of these medications may further diminish respirations and death, already inevitable and imminent, may occur somewhat sooner than it otherwise might, but it will be peaceful. A similar protocol is used for patients who wish to be removed from a ventilator.

### *Nutrition*

As swallowing problems develop, the patient needs to consider whether or not to have a feeding tube placed for feeding. Patients often put off doing this until weight loss is dramatic and every meal a frightening and exhausting battle with choking. The general consensus among ALS patients who have had it done is that they only wish they had done it sooner. Improved nutrition and fluid intake results in a big improvement in their general condition.

Patients whose respiratory status is deteriorating need to consider having the tube placed even if swallowing is not yet a major problem. Once FVC falls below 50%, the procedure for placing the feeding tube is complicated by the patients poor respiratory status. Weakness of the diaphragm (the muscle between the lungs and stomach) and shallow breathing have also been reported to allow the stomach to shift upward further under the rib cage making tube placement difficult. It is not at all uncommon for the procedure to be tried and canceled if left until breathing is significantly impaired. Earlier insertion of the feeding tube is now recommended in order to avoid both weight loss and problems in placing it. The patient can continue to enjoy eating orally and gradually supplement oral intake with tube feeding as swallowing deteriorates. Once placed, care of the tube fed ALS patient does not differ from other patients with tube feedings.

## *Bowel and Bladder*

Although ALS is not proven to directly affect digestion and bowel motility, constipation is a frequent and often severe problem. The ALS patient's weak abdominal and chest muscles and difficulty taking or holding a deep breath make it hard to bear down and supply the push needed to have a bowel movement.

In addition to attention to a bowel program addressing diet, fluids, regular timing, careful selection of pain medications (non-constipating meds for frequent or regular use), and judicious use of laxatives, the care giver can help improve bowel function by providing optimal positioning. Contrary to the design of most toilets and commodes and especially high rise toilets and commodes designed to make lifting the patient to a standing position easier, the optimal position for a bowel movement is a squat -- fanny low, knees high. Any parent of a diapered toddler recognizes the position! A squatting position gives the best mechanical advantage to the muscles of the pelvic area and abdomen to supply extra force in pushing. Placing a footstool under the feet of the patient can help. With a safety belt on to prevent falling, the patient can lean forward on a pillow placed on his lap to further increase intra-abdominal pressure.

Comfort is important in order to concentrate on the job at hand. ALS patients have no loss of sensation and atrophied muscles in the buttocks offer little padding on a hard toilet seat. Most are reasonably comfortable but a bad fit between backside bones and the seat is really painful and may necessitate a different one, possibly cushioned.

## *Skin Care*

Pressure sores are less common in ALS patients than in other immobilized patients. Some textbooks and articles suggest that there are changes in the skin that account for this, but one obvious reason is that ALS patients have full sensation. Pressure areas become very painful before breaking down. As long as the patient can communicate, pressure areas are unlikely to be allowed to reach the point of breaking down. Pressure relief cushions, mattresses, elbow pads, etc., are needed early in the course of the disease. In later stages when the patient is thin from muscle wasting, poorly nourished because of swallowing problems, and unable to communicate discomfort, the risk increases and more frequent repositioning and skin checks are necessary.

## *Eye Care*

As paralysis spreads, the patient may develop weakness of the muscles of the eyelids. Most commonly the problem is an inability to close the eye. This quickly leads to severe drying of the cornea. Redness, itchiness, infection, scarring and vision loss can result. The eyelid does not have to be wide open all the time for this to begin. Early weakness can leave the eyes open just a bit during sleep. A simple observation of the patient during sleep will identify the onset of the problem and allow early intervention --- frequent eye drops while awake and taping the lids closed at night.

Droopy lids are less common but inability to see is another devastating blow to the alert and aware patient who is already dealing with difficulty communicating. Eyeglass frames with "lid crutches" attached can be used during waking hours. Frequent eye drops will be needed to prevent drying.

## *Mouth Care*

Brushing the teeth of an ALS patient with spasticity can be very difficult because the jaw clamps shut in an involuntary spasm. Although it may seem impossible to get anything else into their mouths for cleaning, sponge "lollipops" should never be used with these patients. If the sponge gets pulled off the stick when you are trying to get it out of his mouth, he could choke on it. Instead, use a bite block or make one out of old-fashioned white adhesive tape and sturdy wooden tongue depressors. Stack several tongue depressors together and wrap adhesive tape thickly around one end (half way up the stick) in layers. Make sure that it is securely taped down so it won't slide off. When finished, the taped end needs to be 1/2 to 3/4 of an inch thick. The layered tongue depressors give it strength and the adhesive tape hold them together and pads it.

To do mouth care, you will have to "sneak up" on those hyper-reflexive jaws. You have to get the bite block in place before the jaw clamps down. Try doing it when the patient is relaxed, even snoozing or yawning. (If spasticity is severe, this will be a job that you will have to do when the opportunity strikes, and not necessarily as part of his morning bath.) Quickly put the taped end of the bite block to the back and side of his mouth between his molars. Do not turn it on edge, just put it in flat. \*\*\*\*\*DO NOT put it between the front teeth! \*\*\*\*\* The jaw clamping reflex can be strong enough to break front teeth, especially if they are weak to start with. The molars are a flatter surface, much stronger and intended to withstand grinding pressure. The tape will give some padding to the sticks to protect the teeth, and putting the bite block in flat will spread the pressure out evenly over the teeth. If he does get it between his front teeth, just let go and wait for the muscles to relax. Pulling on it will only increase the pressure and keep the jaw tight longer.

With the bite block in place, his jaws will be held far enough apart for you to maneuver the tooth brush around surprisingly well. If you can't, get a smaller tooth brush or make the next bite block thicker. Doing this while the patient is sitting up or turned at least part way on his side and/or using a suction machine while allowing you to use a little more water without choking him. Be careful with the round plastic "wand" of the suction equipment, however. It does not make a good bite block at all! To get the bite block out, just let go and wait for the muscles to relax. Sometimes by the time you are done with a good brushing, the muscles are already relaxing.

If it becomes impossible to even get a bite block in place, there is a nasty little tool called a jaw screw that can be used. It is basically a short, fat, plastic screw. The tip is placed between the molars and as the screw is turned, the jaw is wedged open. This is very hard on the teeth, even the molars, and there is always some risk of breaking them. You also have to be very careful not to catch and grind up the corner of the mouth in it. In short, it is something that should be used only if the jaw is constantly tight so that getting a bite block in is impossible, and then only by someone trained in its use.

Sialorrhea is the correct term for excess salivation and drooling. ALS patients do not produce extra saliva but swallowing problems reveal the surprising amount of saliva we normally produce and swallow without thought. Sialorrhea is understandably distressing to patients and is a problem often under treated because it may take trials of several medications before one is found that provides some relief without undesirable side effects. Often the patient is unwilling to continue the medication long enough for the side effects to lessen.

Medications that can be tried include:

- glycopyrrolate (Robinul))
- Amitriptyline (Elavil)
- benztropine (Cogentin)

trihexyphenidyl hydrochloride (Artane)  
transdermal hyoscine (Scopolamine)  
Atropine

For or thick mucus production associated with sialorrhea, the addition of a beta blocker, such as propranolol (Inderal) or metoprolol (Toprol) may help. External beam irradiation to a single salivary (parotid) gland to reduce saliva production is being used with good success by some specialists. Botox injection of the salivary glands is used with success in some patients. The effect lasts several months.

### *Range of Motion*

As with any immobilized patient, passive range of motion and gentle stretching exercises are important to prevent contractures and frozen joints. Although such exercises serve no purpose in restoring function in the ALS patient, they are very important in preventing pain. With contractures and frozen joints, it becomes very difficult to position the patient comfortably.

### *Comfort*

One of the most common misleading statements about ALS in textbooks is that there is no pain associated with it. Although it is true that for most patients there is no great pain directly due to the disease process, there are a few patients who do experience severe unexplained muscle and joint pain -- and nearly all patients experience significant discomfort at some point. Muscle cramping and spasticity can be very painful, joints ache as muscles weaken. The small discomforts of sitting or lying in one position reach a whole new level of misery when you cannot shift position enough to relieve them.

In ALS patients all the problems experienced by patients paralyzed by stroke or spinal injury have the added dimension of occurring in the presence of full sensation. Long before there is any visible skin breakdown, pressure areas burn and ache fiercely. Foot drop pulls the skin on the top of the foot until it feels like it will tear. Swollen ankles burn. Subluxed shoulders and frozen joints defy any attempt at comfortable positioning. And then there is the unreachable, unscratchable itch . . .

Most of these aches and pains can be minimized with correct and frequent repositioning and range of motion exercises. Even with good care, however, they do tend to become problematic over time but generally can be relieved with simple analgesics (Tylenol, Ibuprofen). End stage patients often have significant discomfort and require stronger medications.

Patients with spasticity can experience severe discomfort. Spasticity is an upper motor neuron problem and is present to some degree in ALS. For some patients, it is minimal, for others extreme. Spasticity can actually be helpful in maintaining function as the rigidity helps replace normal muscle strength, but it causes jerky, hard to control movements. Spasticity causes a tightening of muscles that results in a stiffening of that part of the body in an exaggerated reflex. It is actually triggering both the muscles to flex and the muscles to extend that part of the body at the same time. Spasticity tends to affects larger areas of the body - arm, leg, trunk, neck. All the muscles in the area tighten up and the entire area becomes so tight it hurts. A simple touch can trigger it and it may persist indefinitely.

Medications usually help, but spasticity is sometimes is a very stubborn problem. One consideration in treating spasticity is to find a balance between relieving excessive and painful spasticity



and maintaining a certain level of spasticity which can be helpful by replacing muscle strength. The meds for spasticity are primarily Baclofen and Zanaflex. In 1996 the FDA approved the use of Baclofen delivered directly into the spinal fluid by an implanted pump for the treatment of spasticity due to spinal cord injury and this is now being used with good results on ALS patients with severe spasticity.

Muscle cramps are very common in ALS. They can occur in small muscles or as large "charlie horses" affecting any part of the body -- fingers, hands, neck, jaw as well as arms and legs. The cramping becomes less severe with time because the weakening muscles simply can't work up a good cramp anymore. Quinine seems to be the most effective medication for muscle cramping. A low dose (half of a 260mg tablet) once or twice a day is usually sufficient. Higher doses can cause muscle weakness. Quinine has a very bitter taste so taking crushed tablets requires a lot of pudding! Baclofen is often ordered for muscle cramps but is seldom effective unless spasticity is being mistaken for cramps or triggering them.

Fasciculations (muscle twitchings) are probably due to nerve irritability. They occur in smaller muscle bundles inside large muscle bundles and can be observed as well as felt. Fasciculations are not so much painful as irritating and have been described as feeling like someone is popping corn inside the muscle. They can be incredibly persistent and strong enough to prevent sleep. No medication has been found reliably effective in stopping them but some patients find that a few minutes massage of the area will reduce them.

### *Positioning*

As with any paralyzed patient, correct positioning is important to prevent contractures, skin breakdown, etc. Because the ALS patient has no loss of sensation, good positioning becomes a critical factor in comfort. One hour spent with an arm unsupported is miserable and can result in days of shoulder pain and sleepless nights.

Being confined to a bed is not comfortable and is generally unnecessary if a chair is adapted for the patient. Being able to sit up for several hours a day will have a positive effect on the patient breathing, digestion, skin, musculoskeletal system, and sleep pattern, not to mention his emotional well-being.

Of course the ideal chair is a wheelchair which has been customized to fit the patient. When such a chair is purchased, it will be fitted by OT/PT. However, with the cost of such chairs, all too often the patient is assigned to a "one size fits none" standard issue wheelchair, geri-chair or recliner. Regardless of what type of chair is used it should be customized to the patient and needs to be "assigned" to that patient. OT/PT should be called in to help the nursing staff make adaptations to the chair. Some important adjustments are:

- Size of seat/distance from front to back (knees to tail bone) If this is too large, it either cuts off circulation to the lower legs or causes lower back pain and skin breakdown from excess pressure on the tail bone when leaning back.
- Position/height of neck rest/pillow. When neck weakness causes the head to droop, a different style of head rest and/or a neck brace is needed. There is absolutely no need for any patient to sit with his chin on his chest or shoulder. It is every bit as uncomfortable as it looks!
- Height from seat to floor/foot rests. This is extremely important in reducing swelling of the feet and development of blood clots.

- Height of arm rests. Too high causes shoulder pain and elbow pressure. Too low leads to subluxation of the shoulder. This leads to aching discomfort when sitting and real pain when lying on the side.
- Comfortable cushions. No standard seat cushion provides sufficient pressure relief for someone paralyzed. A ROHO air cushion or gel cushion is needed. Foam cushions are not recommended. They are better than nothing, but no variety of foam cushion (not even the "egg crate" style) provides adequate pressure relief for sitting. When a cushion is added, the height of both the foot rests and the arm rests needs to be readjusted.

A recliner is probably the worst choice for seating. Few have adjustable seat depth or arm rests (which are generally too low). Most importantly, using the recline position is likely to increase swelling of the feet and ankles rather than reduce it. The problem is that the human body was not designed to bear the weight of the leg on either the calf or the heel which is exactly where most recliner foot rests place it. Weight on the calf cuts off circulation and causes foot swelling. Weight on the heel causes a painful pressure area that will in time break down into a pressure sore. The ideal chair would be shaped to the curve of the leg to spread the weight evenly, but most recliners have a large gap between the seat and the foot rest, leaving no support behind the knee and upper calf -- all the weight is on the lower leg and heel. If a recliner must be used, use it as a regular chair. The foot rest should be used only for short periods of time if at all.

When trunk weakness causes slumping to the side, foam wedges that support the trunk are needed. If the knees/ankles turn in or out, a foam wedge between the knees can improve alignment and greatly increase comfort. A lap desk that wraps around to the sides to support the patients arms is not just a positioning aid. If the patient still has any finger dexterity, the support of the elbows and forearms can also greatly prolong the patients ability to use his hands.

Time invested in getting an ALS patient comfortable in bed is time well spent for any care giver. Because the ALS patient has full sensation, the little discomforts created by awkward positioning can make sleep impossible. Those little discomforts quickly graduate to pain when one is alone, unable to sleep, unable to make even the minor adjustments needed to ease the problem. The only hope for getting to sleep is to call the care giver back again and again until all the nit-picky positioning quirks are addressed. Every patient has their own individual "pre-flight checklist" for comfortable sleep, but some of the basic needs are a comfortable mattress, a foot board to keep the weight off the blankets off the feet and reduce foot drop. (Yes, foot drop is miserably uncomfortable!) More common sleep destroyers are bad pillow position, a folded-over ear (incredibly painful after a bit!), heavy or tight blankets that restrict any weak movement the patient has left. A bedside checklist can help when care givers change often.

### *Swelling of legs*

One very common source of discomfort for the ALS patient is the swelling of the feet and ankles. This begins when leg weakness prevents walking because muscle action is needed to help pump the blood back up the legs. When muscle movement is lost, blood pools in the veins. Water leaks from the distended veins out into the surrounding tissue creating the swelling (edema). With repeated episodes of swelling, water seeps into the tissues even more easily. At the same time, the one-way valves that help move blood upward are collapsing from the weight of pooled blood. That damage is permanent and swelling occurs even more readily.

Doctors often prescribe diuretics, but unless the patient has kidney or heart problems this should be the last resort, not the first. Diuretics remove fluid, putting the patient at greater risk of blood clots and don't address the underlying problem of poor blood flow.

First, make certain that when the patient is up in the chair, the distance from the seat to the floor/foot rests is correct. Having the legs "dangle" is a sure-fire way to cause swelling! Put a box/platform under the feet if necessary to make sure that there is minimal pressure at the back of the lower thigh and knee.

Elevating the feet can help but only if it is done properly. The foot rest cannot be just under the calves and heels as that only further impairs circulation and leads to pressure sores on the heels. Putting the feet up without "unfolding" at the hips is very minimally helpful, possibly even detrimental as that bend interferes with the already difficult job of moving blood upward to the heart. Elevating the legs effectively requires lowering the back rest to a reclining position so that the feet are level with or higher than the heart. Keeping the patient in this position defeats the entire purpose of getting the patient out of bed however. Elevating the feet for a short time several times a day or putting the patient back in bed for an hour or so in the afternoon is a much better solution.

Other interventions:

- Muscle activity also helps even if it is only through passive range of motion exercises.
- Although swollen legs feel cold and the impulse is to warm them, heat will only increase the swelling. Many ALS patients find that just a few minutes of sitting with feet by a heater or in hot sunshine will dilate blood vessels and set off the fierce burning pain of extreme swelling.
- Limiting salt intake is often recommended, but again doesn't address the underlying circulation problem. It is probably sufficient to limit indulgence in very salty foods.
- TED (elastic or compression) stockings can help and ALS patients with severe swelling problems are finding that "boots" that inflate and deflate to help pump the blood along work very well.

*Occasionally I run into someone who seems genuinely interested in hearing what it is like to live with a relentless, progressive, untreatable disease such as ALS. I wrote this for one such friend several years ago. Even though my disability has progressed, my emotional responses have changed very little over the years. Other people with ALS may find it a confirmation of what they have experienced and caregivers and friends might find something worthwhile in it too. BE FOREWARNED: This is not a happy, uplifting message and therefore may not be suitable for people who are having a bum day to start with!*

### ***The Attic: The Hidden Losses in ALS***

You asked what it is like to live with ALS so I think it is time to take you on a tour of my attic. I don't usually take people up there, but sometimes I meet someone who really seems to want to know what my life is like, and the attic is a good place to start. It tells a story of what has happened to me. The story isn't so much about knowing I am going to die as about having to go on living until I do. It is about being handicapped but it has nothing to do with handicapped parking spaces or accessible bathrooms. Those are just inconveniences. The attic is all about losses.

Ready? Watch your head -- the door frame is low and the stairs come up under a low roof! Boy, I need to get John up here to stack some of this stuff, it is really piling up. He is so busy now days that when he has to put something away, he just sets it anywhere. I brought a magic marker so we can at least label these boxes while we are up here.

Here is the box I sent up when I retired. Nursing books, the old coffee mug, lab coat, name tag: "D. Huberty, RN, CNRN, Neuroscience Clinical Education Coordinator." Man, I was so relieved to be free of the physical demands of working! Accomplishing only 2 or 3 hours work in an 8-hour day. Frustrated by projects that I thought would really improve nursing care on the Neuro Unit but simply couldn't physically carry out. Losing touch with the reality of the nurses work because it was changing over time and I couldn't pitch in and do any of it. Whew, I don't miss the frustration and exhaustion! I sure miss the people I worked with though. They call and invite me out to unit parties sometimes, but after a year and a half, those calls are getting infrequent. And I miss me. The neuro nursing expert the staff came to when a patient was in trouble, a family upset, a doctor on the rampage. Someone who taught new nurses what they never learned about neuro in school. A productive (if somewhat irreverent at times) committee member. A respected, well-paid professional. That is who I was for 10 years! Label that box "Identity 11/11/95."

Look at this mess! A box of arts and craft supplies. Yarn and paint and wood burning tools. The pattern for the booties I was going to make for my first grandchild. The cross stitch sampler for my daughter's wedding that I had to give up on finishing. Crochet hooks for the set of Christmas Angel tree ornaments I was going make for each of the girls. And a whole stack of woodworking magazines. The bookmarks for all the projects I wanted to try are still in place! I always thought someday I would have the time for this stuff -- just like someday I would get serious about getting in shape and someday we would build our own house and someday we would go to Hawaii and someday I would organize my closets and someday I would take some classes just for fun, not for a degree. Seems like I just got past the stage of saying "When I grow up, I want to . . ." and now there is no tomorrow, not one that holds any of those things anyway. The feeling that gives me is not really sadness or depression -- I am really quite happy on a day to day basis -- it is a simple lack of interest in something just isn't going to affect me. The last year that I was working I found myself disengaging, taking on only short term projects, disinterested in improving my management

skills, not really caring about the hospital's long term goals. Now I feel that way about life in general. It isn't a crying kind of sad feeling, just kind of an empty one. Label that box "All my Tomorrows."

Whoa, this one is old! There is wallpaper and paint in here from the original owners of this house. This stuff has been replaced and its replacement replaced! Throw that out, but save these rolls. These are what is in the bedrooms now and something might need to be patched. Goodness knows it is going to have to last a long time. John hates doing this kind of stuff. The current decor of this house is here to stay no matter how outdated it gets or how sick of it I get! It is all John can do to keep up with the laundry, he doesn't have time for redecorating and we sure can't afford to hire it done. Besides, he just doesn't care about decorating. He doesn't notice if the towels match or any of that. He runs strictly on practicality. If you use a certain frying pan regularly, you don't put it away, you put it back on the range. If that ugly, dirty, green throw pillow is comfortable, it belongs on the couch even if it doesn't match a damn thing in the room. Things like that drive me nuts. I seem to have an over-developed sense of color coordination! But, since he does all the work around here and I can't even get to half the rooms in the house, in a practical sense the house belongs to him now. It isn't really mine anymore. Even the family room where I spend 16 hours a day isn't mine. MY room would be neat and clean! I was never a fanatic housekeeper, but now I am really uncomfortable in a room that dirty and cluttered. I guess that discomfort is more because I can't do much about it, not the mess itself. I try hard not to nag about housework, weighing my need for food, drink, bathroom, repositioning, etc. against the fingernails on the chalkboard kind of feeling that stack of newspapers and junk mail and pizza crusts accumulating on the end table gives me. Gotta prioritize and ration out requests for help or risk mutiny among the galley slaves! Label that box "Environmental Control."

This whole box of clothes really should just go to Goodwill. I don't wear dresses or shorts anymore. My legs get too cold and they look awful anyway. Those long coats and sweaters are a nuisance in a wheel chair -- you have to stand up to put them on. I wear clothes that are easy to get on and off, are comfortable sitting down, and don't need ironing. Fashion is not an issue! No, that doesn't really bother me. I was never big on fashion or particular about clothes. But it is kind of like the thing with the house; it is all for convenience and practicality, not self-expression. I have to wear my hair in a style that John can blow dry quickly and easily. I stopped changing earrings to match my outfits because that was just one more thing John had to do every morning. I was never a fashion fanatic, but I just don't have choices about how I look anymore. Hell, I don't have choices about much of anything. I get up in the morning at 5:30 am because if I don't, I am stuck in bed until someone can get home again to get me up. I eat whatever someone is willing to fix whenever they have time to fix it. When we go out I eat whatever is easier, neater, to eat. Plain hot dogs instead of chili dogs. I go to the bathroom when someone is around to take me. I go shopping when someone has time to drive me. I guess we can label this one "Freedom of Choice."

Now here is an archaeological find! My wedding dress is in this box! Do you suppose this will ever be in style again? The last hoop skirt in captivity! Well, my daughters laugh at the dress, but I tell them it could have been worse. It was 1969 and I could have been married barefoot in a meadow wearing nothing but love beads and flowers! Here are the veil and the garter . . . invitation . . . guest book . . . Just close that box, I don't want to see anymore. Yeah, we are still married. I still have John but it doesn't seem like we have a marriage sometimes. It is more of an arrangement held together out of financial necessity and duty. There have been some big stresses on the marriage in the last few years but we seem to be able to handle the big things. It is the loss of the little stuff that hurts. Like holding hands -- try going for a walk and holding hands with someone in a wheelchair.

Like standing at the sink doing dishes and having him come up behind me and put his arms around me and cop a feel while he talks to me. Like working side by side on the yard work or cleaning the house or waxing the car. Now I just hang around and watch him work. My contribution is a suggestion or two which, coming from a spectator, is not particularly appreciated. We can still have sex, but it is diminished by not only my physical limitations but also by the fact that he has to take care of me. Try feeling amorous about a really out of shape body you have to bathe and shampoo and dress and undress every day. (If that isn't enough to dampen your enthusiasm, try shaving someone's armpits, helping with their tampon and other basics of hygiene and see if you are still interested!) For all that physical intimacy, we seem to be growing further apart. We never had a lot of interests in common. Classic male/female roles. He was into biking and boating and golf and I liked to read and sew and putter around the house. Now I cannot join him in his activities and he has no interest at all in mine. I can't help with the workload of running a household. And I don't even feel entitled to a say in some decisions in our lives. I am not going to be here long term and he has to be allowed to plan for a future without me. Yes, I still have a husband who loves me enough to stay in spite of it all, but we aren't really partners, playmates, or lovers anymore. So what the heck do we label this one? Let's call it "My Kind of Love."

Hmm, what is in this box? Oh!! No, don't look! Here, give me the tape. This one is stuff that is not for anyone else to see. No, I don't have any deep, dark secrets! It's just stuff I don't want to have to explain. I don't have much privacy in my life now, so I am keeping my past to myself. I don't mean just physical privacy. It is an invasion of privacy to need someone else dress you, help you to the bathroom, and all that, but it isn't so much what people see as what they know. I can't hide a stash of chocolate, try smoking pot, spend money, buy a present, try a new hairdo, read a book, change my clothes, or put a tape in the VCR without somebody knowing. 99% of the time it is no big deal, but I would like to be able to read "Final Exit" without anyone knowing, re-watch the Beatles Anthology without my family wondering if I have crossed that thin line between fan and pathetic nut case, and toss out a whole stack of misprinted pages from the computer without anyone knowing I screwed up! There, it is sealed shut. Just label it "Privacy."

Well, enough of this. Stashed away up in this attic are my identity, all my tomorrows, my control over my own home, my freedom to make choices and come and go as I please and when I please, my ideas of the marriage and kind of love I hoped to have, and my privacy. All lost to ALS. Someone in the ALS group once remarked that the ongoing nature of the losses is what makes ALS so hard to deal with. It isn't like an auto accident where you come out paralyzed. That is a huge loss to adapt to, but people do adapt and go on with their lives. With ALS, you no more than adapt to the loss of one function when you find you are losing yet another. Losing the physical ability is only the tip of the iceberg. You lose so much more. The attic gets more and more crowded.

Thanks for listening,  
Diane

## *Advances "Around" ALS*

One of the frustrations often expressed by those whose lives are affected by ALS is that no progress has been made in the fight against it. I believe that they are aware of the great progress made in understanding the disease process, the biochemical processes at the cellular level, the genetic, genome, and chromosomal tracking of the disease. I believe that what they are referring to is that all of this work has not led to a treatment or cure, to anything that can make a real, practical, day to day difference in our lives. Dedicated researchers have been kissing a lot of medicinal frogs but haven't found a Prince. Rilutek rates as a "Sir" for its effectiveness, or perhaps a "Lord" by virtue of being the first drug with proven effectiveness, but sadly it is no Prince. Radicava is the latest drug. The restriction of it being for newly diagnosed patients without respiratory involvement limits its use and insurance coverage and the horrendous cost is beyond affordable for nearly all patients. It requires repeated IV infusions. It is frequently not tolerated by patients and some report increased weakness. Some have improvement but it remains to be seen if improvement is sustained much less increased. Do I sound skeptical? Definitely. We have made progress on the right to try new drugs but weakening of the FDA's approval process have also left us able to get new, very expensive drugs whose effectiveness and safety is questionable. Well, at least Big Pharma continues to try to develop helpful drugs. The haven't given up on us.

However, if advances "in" ALS aren't yet affecting our lives, advances "around" ALS certainly do. I am referring to technological advances that have prolonged and improved the quality of our lives. This technology isn't something that a person with a falling off a cliff kind of progression has time to appreciate, but my progression has been a long slow tumble down a rocky slope. Over my twenty-nine years of living with ALS, I have seen so many technological advances, each of which I use every day and none of which I take for granted because I have seen life with ALS without them.

Even before I was diagnosed with ALS, I had decided that I would never want to live if living meant being paralyzed and on a ventilator. I was an Intensive Care nurse and dealt daily with patients in that situation. Trying to read the lips of those who couldn't speak, or worse, the eyes of those with facial paralysis, knowing they were desperate to communicate basic needs was frustrating and heartbreaking. An itch they couldn't scratch or an uncomfortable position they couldn't change would be bad enough, but what if they were trying to tell me of a throbbing toothache, gall bladder attack, chest pain? Making their lives physically comfortable was so difficult. Making them emotionally comfortable seemed impossible. Their days were spent staring at the same four walls and a television whose channel they couldn't change. I wondered how they could bear the endless hours. No jaunts down the hall much less a wheelchair ride outside. Just getting them into a chair was an effort because they were tethered to a machine bigger than a dishwasher. Being at home would certainly be an improvement but still, I hoped that their brains could somehow shut down imagination and desire, restlessness and boredom.

When I learned I faced all that because I had ALS, I was horrified. I knew I could refuse to be put on a vent but would I be brave enough for that? Would I even be able to cope with the paralysis before the vent was needed? I couldn't see how.

But that was in 1985. Today I am a vent dependent quadriplegic and quite a happy person. The future I thought I faced in 1985 is far different than the life I lead today! Even before I left ICU nursing in 1987, technology was making changes.

In the late 80s, there was talk of a new type of ventilator that delivered air through a tightly fitted mask over the nose rather than a tube inserted through a hole in the throat. Little did I imagine that first CPAP machine would acquire computer chips by the early 90s and become a Bi-level Ventilator . Bi-level ventilators didn't exist just 20 years ago and were not in common usage until about 10 years ago but today is part of the Standard of Practice for ALS. That bit of technology kept me alive and breathing comfortably for 6 years until an invasive ventilator was needed.

Neither did I envision what that ventilator would be like. In 1985 ventilators were the size of a dishwasher. By the late 80s they were the size of a large microwave. It was certainly not worthy of the designation "portable", but it was a start. The vent of today is no longer the size of a kitchen appliance. It is the size of a laptop computer or even a lunch box and hangs on the back of a wheel chair, making it -- and me -- portable far beyond the four walls of 1985. I can take a jaunt around the yard or neighborhood, a trek to the mall, or trip across the country.

Not all the technological advances have been in medical equipment. A memory foam mattress pad, an great comfort improvement over the "egg crate" foam of the past. My day begins on my Air cell mattress that is even better than any foam. Mattresses that alternate air pressure to relieve pressure points are the next step up. The newest mattresses turn you gently side to side and you set how often, how fast, and how far. I move to my power wheelchair where I sit on a glorified whoopee cushion. The technology behind pressure relief cushions has taken them to new levels of comfort and pressure sore prevention. I can spend a 16 hour day sitting up, something impossible on a standard chair cushion. The power wheelchair itself, although not a new invention, has benefited from technology and is now moved, tilted, reclined, raised, and adjusted with computer technology that gives a greater level of independent operation that is continually being improved upon.

Equipment to move to and from the wheelchair is no longer limited to a Hoyer lift. Ceiling mount lifts are far more convenient and can even move you into the bathroom and the shower or tub. Free standing overhead lifts don't require any construction or holes in the walls. And the slings for lifts are a huge leap forward from the big monsters that required major effort to get under the patient and only might have a hole to go to the bathroom through. Today's slings can be taken off and put back on in bed or in the chair and U shaped design allows for easy use of a toilet, commode or bedpan.

In the bathroom I have another little gizmo that, although available for decades, has improved greatly in 20 years. The electric toothbrush is now battery operated, rechargeable, and as of 1987, has a much more effective rotary brushing direction. Not an earth shattering breakthrough but definitely one that kept me dentally independent much longer and now makes my care easier. Although I don't have one, many people with ALS use heated and padded toilet seats with bidet spray bottom washers and warm air dryers!

Throughout the day I am surrounded by other little gadgets that make my life easier. In the 1990s, when semiconductors for emitting and receiving infrared radiation were developed, remote controls gradually switched to that technology and the proliferation of remotes began. Remote controls are not just for TVs anymore! Stereo systems, VCRs, garage doors and car doors have all gone remote -- and the remote craze hasn't ended there. Heaters, fans, gas fireplace logs, lights, window shades, doors -- nearly everything can be put on a remote. Can't keep track of all the remotes? Get a single programmable remote and teach it to do everything. Can't handle those tiny little remote buttons? Get a remote with over size buttons. Can't handle buttons at all? Get a light



pressure switch and scanning device or get it all available on your computer with eye tracking to make it hands free.

Safety has improved as well. Cell phones make family members reachable wherever they go. Emergency call buttons summon help if we have fallen and can't get up or just need an itch scratched. Chairs with lift seats reduce the risk of falls.

I end my day with a favorite invention that hit the market in the mid 80's. The simple invention of CDs has been a daily joy for me. I love to read and when I could no longer turn book pages I began listening to books on tape -- and dealing with hissing, snarling tapes and the headaches of rewind and fast forward and buttons I couldn't push. CDs are a major improvement. Remote controlled multi disc players make books and music accessible far longer than the tape deck of the 80's. Libraries have books on CD's for listening or downloadable books for reading on a computer or iPad.

Gadgets like these simplify many things for the paralyzed, but for improvement in the quality of life nothing tops the computer. I use it as a remote to control the TV, lights and anything else I can plug into an X-10 outlet. I can use it to answer the phone, to speak words for me when my trach is cuffed and I can't talk, to play my favorite music or movies. And all of this is done with glorious, delicious independence!

Beyond being an environmental control unit, my computer saves me from my worst fear, the mind numbing isolation and boredom, the need to shut down my brain to preserve my sanity. I am occupied, distracted, entertained, informed, and even productive thanks to my computer. Aside from the time-passing simple entertainment of computer games, I use my computer to maintain records for our neighborhood association, retouch, restore, and print photos, design and print flyers and cards, t-shirts, family photo CDs. I can't wield a paint brush or hammer but I have planned projects from simply rearranging rooms to kitchen remodeling, deck design, and landscaping. Being able to see how something will look before beginning has really helped overcome my hammer wielder's resistance to doing things my way. Power in decision making!

Not only have computers come a long way from the basic functions of my Apple IIe of the early 80s, handicapped access to computers has evolved as well. Today I use Click'n'Type, and Dasher, two different type of onscreen keyboards, to type without using my keyboard. Point'n'Click pushes the mouse buttons for me -- all for free. As my ability to move the mouse to move the cursor deteriorates, I can go to a device that moves the cursor by following slight movements of my head. A step up from that are devices that track eye movements to move the mouse. The very latest technology is a page right out of science fiction. Computers can now be operated by thought! This technology is in very early development but is already being used and holds promise for the future.

I had to stop working in 1995, coincidentally the same year that AOL gave access to the new and developing World Wide Web. I had dabbled with the Internet previously but it wasn't until the Web arrived that I found being online to be useful. In just a few years the Web exploded from a few bulletin boards to a massive source of information, entertainment, communication, and commerce. Today I read newspaper and magazine articles (replacing the printed versions I can't manage), use the Internet as a giant encyclopedia to research any topic, comparison shop and sometimes buy, select books on CD to be reserved or renewed for me at my local library, keep in touch with family and friends via email, and keep informed about ALS and communicate daily with others with ALS.

The greatest reward I get from my computer and the Internet is that it isn't a one-way street. I can give as well as receive. My involvement with ALS groups online lets me share my hard won knowledge of ALS with others. I have been able to put up a personal website to relay my nursing information about ALS. I get great personal satisfaction from this technological ability to help

others and have even made my first venture into designing a commercial website. (The heck with personal satisfaction -- I could even make money designing websites!)

Speaking of money, obviously all these tech gadgets aren't free. The medically necessary ones are covered by insurance and Medicare, but the convenience ones are not. Just as when grocery shopping, I compare prices, wait for sales, look for used equipment locally and on eBay. Having had 20 years to accumulate my "toys" -- and upgrade them! -- has certainly been a perk not many with ALS have, but I have learned that the expense is far outweighed by the quality of life these things give me and the help they give my caregivers.

There isn't a day that goes by that I don't use several things that were not available when I was diagnosed with ALS in 1985, and not a day that I don't think how lucky I am to have ALS today instead of before technology contributed to these advances "around" ALS.