The ALS Project

Dedicated to creating awareness throughout the world

This document will help you understand ALS and the people this disease affects.

A few years ago, the famous **Ice Bucket Challenge raised more than $100 million** for the purpose of research, raising awareness, and helping those afflicted by this fatal disease. Sadly, many people participating in the Ice Bucket Challenge had no knowledge of the disastrous nature of ALS or the tremendous physical, emotional, and financial burden this disease places on its victims, family members, and other caregivers who step up to help.

Some ALS facts

ALS, or amyotrophic lateral sclerosis, is a progressive neurodegenerative disease that affects nerve cells in the brain and the spinal cord. The progressive degeneration of the motor neurons in ALS eventually leads to their death. When the motor neurons die, the ability of the brain to initiate and control muscle movement is lost. With voluntary muscle action progressively affected, people may lose the ability to speak, eat, move and breathe.

The loss of the ability to breathe is the major cause of death in a PALS (Person with ALS).

There are approximately 30,000 people living with ALS in the United States at any given time; the number worldwide is estimated at around 450,000. These numbers are hard to estimate accurately because the disease takes time to diagnose. Accurate diagnosis is somewhat dependent upon the type of insurance and financial means a patient has at his disposal, and the fact that availability and quality of medical care for neurological diseases varies from country to country and from region to region within countries. One of the reasons for the low number of people living with ALS is that the disease usually kills its victims within two to five years after diagnosis.

Most of the time ALS is not inherited. In about 90 percent of cases, the person diagnosed is the only member of the family with the disease. These cases are called “sporadic ALS.” The cause of sporadic ALS is not well understood, but may be due to a combination of environmental and genetic risk factors.

About 10 percent of cases are considered “familial ALS” (FALS). In these cases, more than one person in the family has ALS. Most FALS families have many relatives affected. Some can trace the disease back 200 years in their family tree. People with FALS often start having symptoms at earlier ages than in sporadic ALS. Most familial ALS is inherited in an “autosomal dominant” fashion. This means if one parent carries the gene, then there is a 50 percent chance of passing the gene onto one’s children. There are a number of genetic defects that can cause FALS. The most common are c9orf72 and SOD1. There are currently tests to identify the particular genetic defect for about 60 percent of cases. Choosing whether to be tested if you know your family's defect is one of the many hard decisions FALS family members make as there is no current way to prevent the disease and no way to know when it will strike.

ALS typically begins in a limb then spreads to other parts of the body. Bulbar onset ALS starts by attacking the muscles that control swallowing and speech. A less common onset starts with respiration, attacking the diaphragm and other muscles needed to breathe.

Pseudobulbar affect (PBA) is a symptom that affects 15 percent to 45 percent of people with ALS, especially those with bulbar onset. It happens when ALS or certain other neurological diseases or injuries damage the area of the brain responsible for controlling what we typically consider as the normal expression of emotion. The damage can affect brain signaling, short circuiting the normal system and causing episodes of involuntary crying or laughing.

ALS and pain

Even though ALS is a disease that affects motor neurons, it is not without pain throughout the course of the disease. Pain is often something overlooked by the medical community when dealing with people with ALS. One of the most common types of pain that can come early in the disease is cramping. A muscle cramp is a sudden and involuntary

contraction of a muscle. If you've ever been awakened in the night or stopped in your tracks by a sudden charley horse, you know that muscle cramps can cause severe pain. Some people with ALS experience severe cramping in various muscles.

Another type of pain in ALS is caused by spasticity. Spasticity is a condition in which there is an abnormal increase in muscle tone or stiffness of muscle, which can interfere with movement and speech, and can be associated with discomfort and pain. Spasticity is usually caused by damage to nerve pathways within the brain or spinal cord that control muscle movement. Symptoms may include *hypertonicity* (increased muscle tone); *clonus* (a series of rapid muscle contractions); exaggerated, deep tendon reflexes; involuntary crossing of the legs; and *contractures* (fixed joints.) The degree of spasticity varies from mild muscle stiffness to severe, painful, and uncontrollable.

Pain in ALS can also come in the form of a secondary symptom, related to reduced mobility, abnormal stresses on the musculoskeletal system caused by weak musculature, or difficulties in the range of motion of a joint — also known as an articular block. In late-stage ALS, it is very common to have moderate to severe joint pain. The inability to move can also cause bedsores and other pressure sores all over the body.

Treatment

There is no cure for ALS … yet. Currently, there are only two FDA approved drug for ALS. The first drug approved to treat ALS was Riluzole. Clinical trials have shown that it can extend a patient's life three or four months, and in some cases, up to a year. The second drug, approved in 2017, is Radacava. The drug underwent a phase 3 clinical trial in Japan and South Korea where 137 ALS patients were given either Radicava or a placebo. The group given Radicava experienced a 33 percent reduction in the decline of their physical abilities compared to the placebo group. Radicava works by reducing the oxidative stress in the body. People with ALS have high levels of oxidative stress. Radicava is administered via intravenous infusions. Initially, patients have a daily infusion for two weeks and then have two weeks' rest. After that, they need to have 10 consecutive daily infusions followed by two weeks' rest. Each infusion takes around an hour to complete. Reports from patients currently getting the infusions are mixed. It doesn't seem to help everyone with the disease. The clinical trials were performed on PALS with rapid progression and early in the disease.

There are other drugs used to treat the symptoms of ALS including muscle relaxants and pain medicines.

Most strategies for "treating" ALS revolve around providing high quality care and addressing symptom management.

Understanding the degenerative nature of the disease, and undertaking anticipatory planning, is the best strategy for living with ALS as preventing a crisis is critical. Having equipment and strategies in place before they are needed prevents being placed on a waiting list or finding something critical is not available at the time of need.

ALS and the brain

Up to 50 percent of people with ALS will experience some degree of cognitive decline. Some researchers now say the majority will experience some degree of cognitive effect.

The exact symptoms and severity are in direct correlation to the area of the brain involved and the amount of degeneration experienced. Moreover, the effect of ALS upon the ability to breathe and often to take in adequate nutrition are being studied in combination with the brain deterioration.

Behavioral changes relating to ability to juggle complex mental tasks, empathy to others, and anger are common symptoms that can manifest.

For some, the changes affect the language areas of the brain, and the person is unable to find the right words, spell, or give appropriate responses to questions.

When the symptoms are severe, a diagnosis of frontotemporal dementia, or frontotemporal degeneration (FTD) may be given.

For caregivers and loved ones, these changes in the person are often more distressing than the physical degeneration they are witnessing and helping to care for.

What people with ALS would like to let you know

I've visited ALS forums where people with ALS and their caregivers were willing to provide me with suggestions, feedback, and ideas of helpful things to say and do, things that aren't so helpful, and general comments on what life looks like as a person with ALS (PALS) or a caregiver (CALS) of someone with ALS. I've also personally interviewed by phone or in person over a hundred people with ALS and/or their caregivers.

The following comments came from these brave people. Understand that some of them type with one finger, use eye gaze technology to communicate, or some other unconventional method to post on these forums or to send me e-mails. Personal interviews are very difficult because of the emotion, loss of hope, inability to speak, and other issues of the disease. Their caregivers are tired, stressed, worn out, and unsure of what comes next.

- Some of my doctors know more about this than I do. But not many, and virtually no one else. So, if you are a medical professional, consider that, in this situation, the patient living the disease might know as much as or more about it than you do.

- I am not likely to get better. That is a fact accepted by a few people I know but disregarded by the masses. Telling me I look better is certainly a friendly thing to do, but I also know that it is not the truth.

- I have lost a lot of weight. I started out on the chunky side. Now I am described as thin. Quite a few of those around me comment on how great I look because of the weight loss. What they don't understand is how much effort I have put into keeping my weight up where it is.

- Back when I was walking, I often received comments about how slowly I walked. Yep. I walked slowly, but I still walked.
- My voice is weak, but that does not affect my intellect. I am still the same person inside, even though I have difficulty communicating at times.
- My capacity to do things is extremely limited. I have about two good hours on a good day. I want to get the most out of life during those times.
- I know I can still walk very well but you have no idea how hard it is for me to breathe when I try to sleep. ALS progresses in very different ways and starts out in different places.
- I have no intellectual impairment, and I can hear just fine. Please don't shout.
- Yes, I am young. It is rare, but it happens. I worry for my children.

- The little cold or tummy bug you have could kill me.
- My life is not over. I have a lot left to offer. If something is hard to do today, I want to do it. Next week, it may not be possible.
- Please speak to me. I can hear even though I cannot talk. If I need to write the answer to your question on my communication board, please give me the time to do so.
- Talk to me about things. If you want my opinion, please ask. If there is something I can do to help you, please ask.
- If you are a caregiver outside the family (therapist, visiting nurse, hospice care) find out about me and talk with me about things I find interesting or important. Please do this even if I appear unaware.
- Ask me if you can pray for me or with me. Take time to find out my spiritual needs and also the spiritual needs of my caregiver(s). If you feel comfortable in doing so, help support those needs.
- If you're visiting me and find it difficult, try bringing over a movie or offer to read to me.
- I really appreciate leg and foot massages. Even though I am paralyzed, I still have working sensory nerves, so I can still feel everything. Massage also helps circulation and is good for both my health and comfort.
- Try to find time to understand the technology I'm using (breathing devices, power wheelchairs, speech technology for the computer, transfer equipment.) You might be able to help my caregiver and provide her with a break.
- Some of us can be in a lot of pain from muscle spasms, and loss of muscle and fat around nerves and joints. If you are one of our health care providers, please address the needs associated with pain. Decreasing the level of pain can mean the difference between a good day and a horrible day.
- Medical marijuana should be legalized EVERYWHERE for ALS. I have to travel across state lines to get the type of medical marijuana that helps my pain. It doesn't even have much THC in it, so I don't get high.
- If I am unable to meet a commitment, please understand. I am in a lot of pain and, sometimes, I just need to rest.
- When I was first diagnosed, many people offered to help. Please make good on your promise. The more I progress, the more help I will need. There are so many things you can do to help me. Here are just a few: cook and deliver a meal for my caregiver and me; go grocery shopping for me; do a few loads of laundry; help clean the house; drive me to the doctor, to a park or the beach, to church, or to visit someone; take my dog for a walk or to the vet; ask my caregivers what they need.
- Have a listening ear. No advice needed, just listen. You may hear me cry and lament at times or you may hear me laugh. Just sit there and listen while I can still talk. Converse with me when you feel you have the words. When I can't talk anymore, come talk to me. Bring a friend and talk to them in my presence. I may not be part of the conversation speaking-wise, but just to listen to something other than the TV or audiobooks is a good thing. Break apart the monotony of my existence with tales from the outside world. Keep electronics out of it. My world consists of electronics, and I don't want to see things on your phone. I want to see your face, hear your voice, see a live, fully dimensional person instead of a flat-faced, unmoving photographed Facebook face and typed words. I get enough of that. Bring me flowers to look at. Bring your pet for me to see. By all means, bring your baby! I want to see babies. Put their cheek on mine so I can smell their newness. Take me outside, if you can, so I can see the trees and feel the air and listen to the birds.
- Please ask my permission or my caregiver's permission if you plan on bringing someone or something (pet) with you.
- Please don't tell me about magic cures, lotions, and potions. Discussing this with me just uses up my very limited energy, energy we could be using doing something valuable.

- If you knew someone with ALS, please do not assume my journey or rate of progression will be the same. Just because the PALS you knew traveled doesn't mean that I am able to travel. The best way to discover things that will make my life enriched is to ask my caregiver or me.
- Please, please educate yourself on ALS. You can be such a value to me if you understand this disease.
- Each one of us with this disease is different. What might be a critical need for one PALS might not be necessary for another. Take time to understand my needs and know that they will change as I progress.
- Many people with ALS have other health issues that complicate treatment such as diabetes, depression, or heart disease. Some people with ALS cannot tolerate various medicines because of the side effects. Something as simple as constipation can be a medical emergency to a person with ALS.
- If you have a special skill, please make it available to us. ALS is financially devastating, even with the best of health care insurance. My caregiver can only do so much, and it breaks my heart to see her worry so much about my needs while neglecting her own. She needs someone to help her with our money, do light cleaning around our apartment, and cook a few crockpot meals a week.
- I have waited months for a BiPAP machine to help me breathe at night. I am on Medicare with the richest supplement plan money can buy. Over half of my monthly income goes to health insurance premiums, drug co-pays and supplements. The wait on top of the financial burden makes my suffering so much worse. I have no idea how I will be able to afford home health care of any kind. I tried to get long-term care insurance years ago, but was denied due to an insignificant pre-existing condition. If you are reading this and have any influence with legislatures, please help us.
- I am afraid of being abandoned by my family and friends. Some have already left. I feel like I need to walk on eggshells and put on a happy face for people to want to be around me. I feel like I can't complain because I will be considered negative. When people leave, I cry. Please let me be real with you. It's the only way I can get through this disease.
- I'm terrified my caregiver will get sick or leave. The pain in his eyes breaks my heart.
- I can't get comfortable in my chair or in my bed. Sleep is my only relief. I've tried all the muscle relaxants and most of the pain medications. I get furious when I read ALS does not affect sensory nerves, only motor nerves. Constant spasticity and muscle cramps prevent me from enjoying my family.
- Our local ALS Chapter doesn't support the needs of its patients and caregivers. We make a huge effort to get to the meetings, but clueless speakers are brought in to waste our time, and our important questions go unanswered. Why can't someone approach hospitals, doctors, and medical equipment suppliers in our area to organize fundraisers where the money directly benefits the patients? We need to raise funds for in-home help, remodeling to provide a safe place to live, vans that accommodate our chairs, and other necessities to keep living. As a PALS, I cannot do this. My CALS is too busy and worn out that he can't do this. Somebody must be able to help us. Perhaps someone could write a grant.
- If your PALS belongs to a church, please get the congregation involved. Most of the members won't understand what ALS does to a person. Take the lead and help them understand and get involved volunteering. Talk to the priest, pastor, or other religious leader to find out if he or she has suggestions that might help your PALS.
- ALS has affected the part of my brain that controls emotional expression. I may laugh or cry in a way that makes you uncomfortable. I know that my response is inappropriate, but I cannot make my body obey what I am feeling inside. When this happens, I may leave the room or look away from you so that I can regain control. Please ignore these outbursts, as they are quite embarrassing.
- Doing something as simple as making my bed totally exhausts me. Bending over to feed the cats causes my legs and back to go into spasms. I dread routine tasks that, just a few months ago, I did mindlessly.

- One of the most frustrating things for me is that my mind still works, it does not stop. My emotions and body are in turmoil, but my mind works. I want to do something and I just physically cannot. I jump up out of the chair and almost take a face plant because I forget that my legs and hands just don't work that way anymore. It is so frustrating. But I am still a person, I need others to see that my mind works and that they don't need to speak loudly to me or dumb everything down like I can't understand. I need to choose what is best for me and have that choice honored. Unfortunately, it is the medical practitioners who are the worst at this.
- For the first six months after my diagnosis, I was in denial. I was mad and refused to be proactive in addressing my future needs. That was a big mistake. ALS waits on nobody. I had no idea how long it would take to get the medical equipment I need to survive. My motorized chair might take up to six months to get to me. Even then, I will have to pay for part of it. Medicare covers the rest. There are supposedly loaner closets but I have had no luck in finding a loaner chair that didn't give me more pain than I am already in.
- Some PALS are extremely sensitive to loud noises, bright lights, and strong fragrances. Please ask the PALS or CALS if this is the case. When in doubt, do not wear perfume or scented lotion when visiting.
- Most of us deal with extreme fatigue. We might take several naps during the day, even early on in the diagnosis. Please don't take offense if we have to cancel an event. It is sometimes impossible to know when we will have a good day.
- My time is so valuable. If you make a commitment to visit, take me to the doctor, or help me in some way, please try to be on time. I so appreciate anything anyone does for me.

What caregivers of people with ALS would like to let you know

- If it is too uncomfortable for you to ask my PALS about ALS, please ask me.
- If you are a close friend with my PALS, try to keep that bond until the end.
- Remember that the person with this horrible disease is completely aware of what is going on. His or her mood might reflect fear, anger, or hopelessness. Other times, he or she might be happy, relaxed, and friendly. Please don't take offense; just love them.
- When you first find out a friend, co-worker, or loved one has been diagnosed with ALS, telling him you're sorry and that you care about him is most appropriate. A gentle hug, a hand on the shoulder, or another warm gesture will be appreciated. Never underestimate the power of human touch.
- If you offered to help when you first found out about the ALS diagnosis, please continue to ask as the disease progresses and make yourself available to the caregiver. We've all heard, "If there's anything I can do, let me know." We've heard it many times. Please don't make empty promises. I tell everyone who has asked this question that we are fine right now, but the day will come when I will need your help. Follow up with the caregiver frequently.
- I've found that most people are shocked when they learn about my husband's diagnosis, so I overlook the blunders that some people make when I first tell them. When people are told of a disease that is always fatal and has little or no effective treatment, they grasp for something meaningful to say. I realize that these folks have very good intentions. The best thing you can say is, "What can I do to help?" If you feel you cannot help, it's best not to offer.
- I wish all our close friends would research ALS and be prepared to help, even if it is in some small way such as grocery shopping or mowing our lawn. If ten people could donate two hours a week, that would be a blessing. I feel that I'm in over my head, and really need family and friends to step up now.
- Never compare ALS to other diseases. ALS is not cancer. With cancer, there are many treatments that can prolong or even save lives. Not so with ALS.

- Please don't tell my PALS or me how bad we look. We know.
- We've heard all the stories about alternative treatments, and we've tried many of them out of desperation. We have both spent hundreds of hours researching treatments online, in person, through ALS clinics, and communicating with other patients and caregivers. The best thing you can do for us is to find a way to help in our daily struggle.
- The lack of general knowledge by medical professionals about neuromuscular diseases is astounding. We have met emergency room nurses and doctors, primary care physicians, and specialists who have no idea how to treat restrictive airway diseases. I understand that every medical professional cannot know everything, but basic information and the ability to assist should be available.
- My PALS has been cleaned and made presentable for your visit. The before and after can be quite different.
- When you visit our home, please understand that things are organized for my PALS' convenience. Please help me keep things organized in a way that promotes effectiveness and efficiency in helping her.
- It takes a minimum of two hours to get my PALS fed, medicated, showered, and dressed in the morning. Then I need time to clean up and get myself ready. Please understand that early appointments are very difficult for us. If you are visiting, we won't be up and going very early.
- My PALS cannot stand, and I cannot lift him. So we try to schedule our outings around his bowel movements. If we misjudge, he's stuck wearing a dirty diaper, and that is no way to socialize. We try to be on time when we make plans, but sometimes it's just not possible.
- We often cannot predict when my PALS will need to sleep all day. If you've planned a visit, please don't be offended if your visit is with me.
- Depending on the stage of ALS, a PALS might be able to function pretty well or need 24/7 care. The latter requires continually monitoring him or her, adjusting the bed, repositioning arms and legs on the bed, turning to avoid bedsores, adjusting the BiPAP mask, feeding, making transfers or emptying urinals and responding to a variety of other issues while he or she sleeps.
- Many caregivers survive on very little sleep because the PALS' breathing alarm goes off, interventions such as cough assist and suctioning must be done throughout the night, and shifting the PALS' position to avoid bedsores has to be done.
- Please don't let me and other caregivers down. You must be reliable no matter how small the task might be.
- Most CALS don't have time for lengthy chats on the phone. They are too busy trying to tend to constant needs throughout the day. Please don't be offended if I have to cut our call short.
- As a caregiver, the things that have helped me the most are cooking meals, buying groceries, and staying with my PALS while I run out to go to my doctor. Someone organized a meal train for us where various neighbors in our condo took turns bringing over a healthy meal each night for several weeks. Another person helped organize a respite time out for me for three hours, two days a week. I was able to go for a massage, get my hair cut, and work out.
- If you are a nurse or other health care provider, please consider donating an hour or two of your time to help someone with ALS.
- While my PALS is sleeping during the day, I am very busy trying to get caught up while keeping one eye on her. I often eat on the run or while I'm cleaning. If I sound rushed, irritated, or confused, please understand.
- I find it hard not to project my needs and opinions onto my PALS. I'm always second-guessing what she wants or needs, and I now realize it's really what I want or need. Sometimes, I find the doctor talking to me and looking at me instead of her when, really, the choice is hers.

- If hospitalization is necessary, try to stay with your PALS or arrange for someone who is familiar with ALS to stay. Treatments in a hospital that are routine or common can kill a person with ALS.
- "ADA Accessible" doesn't mean it meets your PALS' needs. If your PALS is traveling, make sure the places you stay and visit can accommodate your PALS. Call hotels and ask for specifics such as door widths, ramp availability, and shower set-ups. Climbing into a tub is accessible, but may not be possible.
- I keep a "to-do" list handy at all times. When someone offers to help, I have the list available.
- A PALS needs a team of specialized health care professionals available. Patients also need a binder outlining their treatment plan and the latest changes in their progression. ALS clinics can be exhausting. They need to be restructured to accommodate the needs of the PALS at that given time. A PALS does not need to see everyone at clinic every time. Going to clinic can mean driving long distances, staying in hotels, and dealing with tests, waits, and meals. If you have the time, please offer to help the CALS take the PALS to clinic. Remember to bring snacks and water on clinic days.
- If you are a local doctor treating a PALS, please have numbers available of all the patient's health care providers. If you are the primary care physician, help your patient get a local team together with experience in ALS. Untrained health care professionals can do more harm than good in an emergency. Keep a list of each person on the team's contact number.
- Hygiene is exhausting but necessary. If you are working with an ALS patient, please keep an eye out for any signs of poor hygiene and address it. Also, look for dry skin that doesn't respond to therapeutic body cream, sensory changes, pain, mental anguish, fungal infections, bedsores, boils, and strange smells. These all need to be addressed.
- It WILL be necessary to train caregivers and outside help in how to clean peg tubes, how to transfer patients, and how to operate and adjust other medical equipment. It's best to do this early and have more than one or two people trained.
- The equipment we have breaks down. The power goes off. Unforeseen things happen. Help someone with ALS have plans in place and people in place for these emergencies.
- I contacted my local ALS Chapter representative, and she said there was nothing she could suggest to help me with emergency home care of my husband. The nursing homes in my area will not take ALS patients who use a BiPAP machine, even if they are not vented. I can no longer handle his aggression toward me, and there is nowhere to turn.
- We tell people that the main thing they can do is treat us like US. We need your friendship and understand that you might be intimidated by our situation. Please know how much we appreciate your love, friendship and help.

What Not to Say to a CALS

1. "Let me know if you need anything." A CALS will never let you know. Life is too overwhelming, with a thousand needs, which are a blur when you offer to help. Instead, visit and observe. If you see one pouring a glass of whole milk from a half-gallon carton, keep this in mind the next time you go shopping. Ask if you can drop off a half-gallon carton of whole milk next time you are going by. Don't bring a plastic gallon container of skim milk, thinking it's healthier and twice as much.

2. "You're exhausted. Have you thought about placing your loved one?" CALS get so mired in care that they'll go on beyond the point of exhaustion. Sadly, some die after pushing themselves too hard. Despite this, CALS tend to think they can manage. When you walk in a CALS's shoes, you'll understand. It is far better to support the overwhelmed

CALS's choice of life and help out. If helping frightens you, you could offer to help pay for a day of adult day services. Every little gesture adds up and means a lot to an exhausted caregiver.

3. "Why do you do that when you can do this?" Instead of judging the CALS, try to understand the reasons behind the actions. Most CALS have researched ALS a hundred times more deeply than you have. They have probably tried a bunch of things before they found whatever they are doing now that works. It may not look like the best way to you from the outside, but there may be many very good reasons why certain things are done in certain ways.

4. "You're a saint." This may be how the caregiver interprets this well meant comment: "I'm doing all I can to care for him. I wonder if I'll survive. When I hear that I'm a saint, it puts pressure on me to try harder."

Caregivers give until they can't give any more, and no one wants to add any more pressure.

It's the little things that add up and overwhelm. Yet, helping with the small details will only take a few minutes out of your day or a couple of hours each month and be greatly appreciated by the stressed-out, overwhelmed caregiver.

True stories written by people with ALS.

All names have been changed.

John's story

The experts and different websites all tell us that the malfunctioning motor neurons and subsequent muscle wasting is not painful. Technically the loss of motor neurons is not painful; however, ALS can be a very painful disease.

I have excruciating muscle cramps in every imaginable part of my body. This morning, I woke up to my left forearm cramping so badly that it caused my fingers to curl and lock. While this is not the motor neurons dying or the muscles wasting away, it is caused by ALS. What little muscles I have left in my arm have to do extra work to just function during the day. That, with the work that the fasciculations constantly put on them, cause them to fatigue and cramp. The worst for me is when my neck and jaw cramp and lock up.

I have extreme headaches. These headaches are most likely caused by CO2 building up in the blood during sleep. This is caused by our breathing muscles not properly working and their inability to expel carbon dioxide. Again it is not the motor neurons acting out or the muscles dying, but it is a direct result of these things happening that causes our diaphragm to no longer adequately function, leading to the CO2 buildup and subsequent headaches.

Last week, the muscle on the top of my left foot was in extreme pain. This was caused by the other muscles around my foot weakening and being dead, putting more strain on the small muscle at the top of my foot. This forces a muscle not meant to do the work to compensate for the muscle that is paralyzed. Again, this is not pain from motor neurons dying, or muscles dying, but it is caused by those things happening leaving our bodies in a weakened state.

I go through coughing and choking fits. I do not know why my body secretes so much phlegm, but it does. It is in some way a direct effect of this monster.

The anguish my mind goes through with my own pain and inability to function pales in comparison to watching my loved ones suffer watching me waste away. Seeing the look on my daughter's face and listening to her explain what is happening to me to her children is unbearable.

Despite my suffering, financial ruin, and fear, the Lord will see me through this horror. With God's grace and mercy, I will continue this fight in order to serve my family as best I can. I will not let ALS take away my love for them by turning me into a mean and bitter man. I cannot stop ALS, but I can define who I am and what my actions will be.

Amy's Story

I guess this disease doesn't discriminate. I was always careful about my health. Thin, muscular, athletic, never smoked, never drank, always ate clean, took supplements, exercised every day and had a positive attitude. It got me anyway.

I'm an attorney. Worked hard, took nice vacations, loved life, had great friends. It got me anyway.

I'm a mom. Loved my kids, cooked healthy meals, went to games, taught them well. It got me anyway.

This disease isn't supposed to strike the young. It did.

There is no cure. None. I live in the United States of America. Our health care system is lacking so much. My entire retirement is being spent on this disease. I have to move. I have to relocate my kids. I have to make difficult choices about how and when I will die. I have to tell my kids why I can't hold them. I have to watch my friends look at me

with pity or not come around at all. I have to explain over and over to every new health care worker my needs, my limitations, my disease, my wishes.

This is just the beginning.

Kevin's Story

This disease is taking me both physically and mentally. I laugh and cry inappropriately. I'm ashamed and embarrassed because my buddies look dumbfounded when this happens. Something as simple as watching a football game with my brother has become impossible. It took the doctors two years to figure out what was happening to me. I thought those two years would be the worst two years of my life, but I was wrong. Every day is worse than the day before. I can't get control of my anger, and it feels like ALS is something I deserve. I know that doesn't make sense, but it haunts me.

The VA helps with equipment and counseling. They claim I have PTSD and ALS. I was told not to go on hospice until I had all my equipment. I can't get used to the Trilogy at night. They told me I needed to get a feeding tube soon before my breathing gets worse. There are so many decisions to make, but I had to give my brother permission to make decisions for me because of my mental condition which they told me is part of the disease.

In the beginning, the hardest part was listening to my buddies talk about all the things they were doing with their families and friends. Then I was conned into trying all these different kinds of "cures." I think some might have had good intentions, but others were out to get what little money I had. Now I just shut them down, shut them up, or shut them out. I might have a brain disease, and I might even act inappropriately, but I can still recognize self-absorbed (expletives deleted) when I see them. My advice to anyone with this disease is to keep your options open and watch your wallet.

Ruth's Story

When I first went to my family doctor, he told me I was experiencing stress from work. The twitches in my right foot and leg were common, and "everybody had them." I tried to explain that I was tripping, and he got irritated and told me to start wearing sneakers instead of shoes. A month later, I was limping and just low on energy. I went back, and he gave me a prescription to see a physical therapist to build my leg muscles back up. I spent several months doing exercises and stretches, but they only made my leg weaker and twitch like crazy. The physical therapist thought I wasn't trying hard enough or doing the exercise "homework" between sessions. My doctor reluctantly ordered an MRI of my lumbar spine which was normal. "See, I told you nothing was wrong." By then I had a hard time climbing stairs, and my left leg joined the party. I was also starting to get cramps and felt like my balance was off. I frequently ran into walls or misjudged stairs.

Nine months later, I found myself in an ALS Center of Excellence, listening to the doctor confirm my diagnosis. He suggested that the weight lifting might have accelerated the progression. It didn't matter. Nothing would have changed the outcome.

Today, I cannot walk. I cannot feed myself. I live in a motorized wheelchair 12 hours a day and a bed the other 12 hours. I am being fed through a tube, but can still talk and eat certain food. I'm in constant pain. My niece and her daughter take care of me. I try very hard to have a positive outlook and not be demanding. My niece hired a CNA three days a week, and he is very nice to me. He washes me, reads to me, dresses me, and even makes me laugh. I had to give up my cat because my niece is allergic, but my best friend took her and brings her over in a carrier so I can visit with her outside the house. I never thought I'd be so excited when I heard I qualified for Hospice. Now, my niece will receive help every day. I also got accelerated benefits from my life insurance, so we can buy a van that is wheelchair accessible. I think I have it better than most people with this disease, and the reason I agreed to share my

story is so people would know how much we need to fund research. ALS is not incurable. It is underfunded. As many people get diagnosed with ALS as get diagnosed with MS. It is not as rare as people think. You just don't think about it until you or someone you love gets diagnosed.

Murphy's Story

I think the psychological stress is the worst part of ALS. At least it is for me. Maybe I'll change my tune later on when I can't move. I can still function with minimal assistance, and I'm online most of the day searching for something, anything that might slow my progression. I guess I had a lot of the so-called risk factors. I'm a veteran, I smoked, I had a couple of bumps to my head playing contact sports. I also worked with a lot of pesticides and chemicals after I left the Army.

I agreed to write my history on one condition, and that is so I could tell everyone the ONLY thing that has helped my symptoms is marijuana. It isn't legal in my state so I have to roll the dice in hopes of getting a good supplier and good product. It helped me gain back all the weight I lost, helped me to get restful sleep again, relieved my anxiety a bit, and even helped the wicked cramps in my feet and legs. Now, I'm faced with probably having to move to Colorado or Washington where I can get the exact kind of medicinal marijuana that helps me. The other issue is that smoking it is getting harder as I lose my ability to breathe good, so I'll need drops or candy or some other delivery method that doesn't affect my lungs.

I got diagnosed pretty fast compared to the people I talked with and the horror stories I've read online. I went in for what I thought was a pulled hamstring, and, after the examination, I was referred to a neurologist. The neurologist did an EMG and referred me to a neuromuscular neurologist, and I was diagnosed. The total time between my first visit and diagnosis was less than six months. The specialist gave me a prescription for Riluzole and asked me to come back in three months.

I'm divorced with no children, but have some cousins out near Seattle who want to help me. I have to make a lot of decisions, and I hate selling my house and moving away from my friends, but I'm lucky to have relatives in a state that can supply me with the only thing that has helped, so far. The VA has been good to me here. I have no complaints. I have a good monthly income and am planning a trip to Seattle this summer to check everything out and spend some time with my cousins.

Tess's Story

We are a FALS family from California. In my lifetime, I have lost my mother, three of her siblings, and my brother to ALS. My mother was one of five children, and only one survives. My uncle has lost all his siblings and one nephew so far. A niece (me) currently has ALS and at least one more niece carries the gene. In all likelihood, she will get ALS. Four other nieces and nephews are at genetic risk.

In the last 26 years, there have been less than three years that at least one family member did not have ALS. When my mom died, my brother was diagnosed one WEEK later.

When you are FALS, as soon as you know you have ALS, you can see the whole road ahead. There is no learning curve, no denial; it is just there. The moment I found out I knew. It was like a lightning bolt. That was last winter.

Being FALS means that when you hear your cousin has a hand problem you immediately think the worst and it is not being a hypochondriac.

Being FALS means wondering about the children you have and worrying about future children. How can you risk passing this along? Surely there will be a cure for them...but that is what our parents thought.

We take the long view -- we have to. I belong to a Facebook group, and some of us have participated in several research studies each hoping to help find answers. My head knew better, but my heart thought if I did just one more test or gave one more tube of blood, I could save my brother. Of course, I was wrong. Now I am fighting for our children.

True stories written by caregivers of people with ALS

Julia's Story

I feel like I'm living in a nightmare. My fiancé was diagnosed with ALS a few months after our engagement. One day, we were planning our wedding, and the next we were fighting his insurance company to pay for a second opinion. Needless to say, there will be no wedding because he has very little money saved and, most likely, will declare bankruptcy. I have more saved, and my car and house are paid off, so it wouldn't be wise to combine our assets at this point. The hardest part, by far, is the lack of help we've had in the way of navigating the system to get the things he needs to help him survive each day. Right now, he can still walk, and I still work full time. I know both of these are temporary because I'm his only caregiver. He has one sister, but she lives far from us. She has come to visit several times, but she works full time, too. She can only stay a week but she is helping us financially. We're both grateful for her help.

I can tell you that I'm so stressed out about our future that I'm having panic attacks, and so is Tim. His insurance has a huge co-pay for counseling, so we are using mine to get help. Tim is more worried about me because he thinks he is a burden, and, when things get worse, he doesn't think I'll be able to handle it. Our counselor is helping us with a life plan, and we're supposed to have goals but, at the same time, make each day count. How do we even do that?

One day, I feel angry, the next helpless. We go to the local ALS support group, and looking at the PALS who are more progressed than Tim makes me cry. I don't understand the lack of home help when you're on Medicare and have this disease. That's what makes me angry. It would cost the government much more if Tim had to go into a nursing home. I won't let that happen.

Dee's Story

ALS is a thief that will still voices. All voices. Voices that were lyrical and smooth. Voices that were loud and boisterous. Voices that once were used to laughingly call their children's names, or to whisper sweet nothings into the ears of a loved one. Voices that were used to tease friends or calm fears. Voices that shared memories of the past and voices that shared dreams for the future.

ALS will quiet all those voices until they are no more.

Today, Mike used his Eye Gaze computer program for several hours, typing each letter, one gaze at a time, to form the words his voice once so freely spoke. He wanted to share some stories of his childhood with me.

I was looking forward to reading it because he is able to share more using his computer. It's difficult to have long conversations using our alphabet card. His eyes get heavy, and I have trouble seeing when he is blinking on a letter. And holding the card at an angle that allows him to see means twisting and leaning, which strains my back. Even more challenging is to remember the beginning of the sentence by the time you get to the end, (we do a lot of "start overs"), but, in the end, the best way is to write each letter down as he blinks on it. I don't know how he doesn't get more frustrated with the whole process.

Today though, he was using his computer voice to share. His concentration is amazing, and he was so focused. Click. Click. Click. Click. For hours. And then he typed out a very short sentence and clicked on the "speak" button ...

"I lost it all."

Sigh. This has happened before. The program isn't like a regular word processing program, and he hasn't got the saving down pat. We turned the computer off, wiped his eyes that tear and burn from the intense gazing, and settled down for a nap. The story will have to wait for another day.

We all need to cherish our ability to speak. We need to choose our words carefully and make them matter. Once spoken, they cannot be taken back. But if not spoken, those thoughts may be forever lost.

We need to say “I love you” to all those we love as often as we can.

We need to use our voices to do good and to teach our children to be good.

We need to sing out loud and laugh freely and often.

We need to use our voices to give comfort and to help those who need it.

We need to use our voices to make a difference in this world.

ALS patients need others to be their voices to find a cure. They need YOUR voice to speak loudly for their voices that will soon be or are now silent.

The Ice Bucket Challenge brought us out of the hidden corners where we were tucked away unnoticed. Please consider using your voice to help us find a CURE.

Ray’s Story

I know we need to find a cure for ALS. I hope we find one soon.

What I want to say is that we need help to get through this horrendous disease as it progresses and gradually crushes the lives out of the people we love. We need resources to help us live now. Some of us, myself included, can’t just go out and buy a new house with all the ADA modifications or buy a van so I can drive my wife, who is confined to a wheelchair, to her doctor. I’m a hard- working husband. My wife had a job until she could no longer pick things up. We saved a little money and had enough for a down payment on a modest home in a good neighborhood when my wife got pregnant. We were delighted when my wife delivered our son, Gabe.

Shortly after Gabe’s birth, my wife began having symptoms. We thought that she was just tired from the baby, but that was not the case. Then we found out that she had Type 2 diabetes, which we found surprising because she had always been thin, active, and nobody else in her family had this disease. After that, things went downhill fast, and her doctor sent her to a neurologist. He figured it all out and sent us to an ALS specialist who made the diagnosis. We were horrified because my best friend’s dad died of ALS, and I knew what was ahead of us.

I have no idea how we’re going to get through this disease and give my wife any quality of life. Each day, I have to make impossible choices like do I cash in my 401k to buy a van that will accommodate a motorized wheelchair or do I continue to put Gabe in his car seat and lift my wife from her wheelchair to the passenger seat, fold up her wheelchair, put the passenger seat in full recline to get her comfortable, then stick her wheelchair in the trunk just to get her to the doctor’s office? My wife has to make impossible decisions, too. She has already decided to get a feeding tube, but she doesn’t know if she wants a vent or not. This disease is killing her mentally as well as physically. She doesn’t want to leave Gabe and me, but she doesn’t want Gabe to miss out on all the experiences normal kids get to enjoy.

Last night, I had a dream that my friend's father was still alive. Today I woke up feeling guilty and helpless. Guilty that I didn't do more to help John's dad, and helpless that I can't do enough to help my wife and son.

Rachel's story (newly diagnosed)

As it often does, this thing we face now began with a whisper, not a roar.

In February, Sam had a fall on some steps. In typical Sam fashion, he downplayed the accident but he did begin to say that his knee hurt. As weeks went on, he would say his walk felt different.

In April, he could not raise the toes on his left foot nearly as high as the toes on his right. Drop foot. He realized he needed to go to the doctor.

In May, Sam had an EMG/NCS. It was obviously not good, but we were getting no details. Just to come in for an MRI of the brain, leg, spine with a follow up appointment to "discuss the results." I really feel at this point we deserved - and should have asked for more communication.

That appointment to "discuss test results" was surreal. It was then that a real live MD actually said it was either an "autoimmune condition" or ALS.

When we went back to the doctor for the results in June, we heard that the blood and spinal test had revealed nothing. This was actually very bad news, but the doctor explained that we still did not meet the criteria for ALS. He wanted us to go to the U of M for a second opinion. He said some things here were not consistent with ALS and seemed to think it might be one of the rare motor neuron conditions, or some weird thing usually found in Japanese people.

I came home from work, and Sam showed me something he could no longer do with his left hand that he easily did with his right hand.

Honestly, in some respects our appointment at the U of M was a relief. We knew at this point anyway, and while it was not easy to lose that last shred of hope, finally we had clarity. Dr. Smith spoke so kindly and gently to us. I will always remember that, and I thank him for it. Note: kindness matters, tact matters, and your own frustration at what medicine cannot yet do and does not yet know is not an excuse to tell people they may be terminal with all the care of making a lunch order.

After spending most of our day at the clinic, we celebrated my birthday with friends the following day. We knew that right now, we were living rather than dying with ALS. We booked a vacation, planned for the future some, but did not dwell on it. Today is truly the "present" that we have. Our friends and family were pretty awesome.

There are no words to describe the pain of this, the mourning for all of the years together we will lose to this disease. We have no idea how this will progress, only what will happen in the end. Yet our story is not over, and there are still good times to come.

Katherine's Story

The nightmare began with small things that did not seem connected but slowly worsened. The very first thing I noticed as his wife was a change in his personality, more than a year before the physical symptoms started up. He went from a very upbeat, positive, energetic person, to an irritated, negative, lethargic person.

Jim ran his own café, and his life revolved around food, talking and laughter. His first physical symptoms were choking on water, slurring his speech and being unable to control hysterical laughter.

It was six months before these symptoms were constant and severe enough for him to agree to see a doctor.

We spent the next seven months waiting to see the next specialist or dentist as we were referred on, only to have each new specialist totally mystified while Jim's speech and swallowing worsened, and he sold his café. It was torture for him to be asked if he was drinking, or what was wrong, and have no answers.

His hands also became very weak, and he couldn't open a can or a cardboard packet. He was becoming so fatigued he was closing the café and coming home several hours early.

Finally, we saw a speech pathologist who nailed it within five minutes and said those awful letters – ALS. Within a month, we had a formal diagnosis at an ALS clinic, and our lives were turned completely inside out.

Jim was rapid progression as is often the case with the combination of Bulbar onset ALS with FTD (frontotemporal degeneration). In just 11 months from diagnosis, he lost the use of his arms, he fell often, and twice sustained serious injuries that impacted on his quality of life severely.

He did agree to a peg for nutrition, but waited until he was emaciated and dehydrated, and he never adjusted to taking in enough nutrition to stop losing weight.

He never agreed to a BiPAP. He was still doing some standing transfers during the day, but could not walk, and was hoisted for many transfers and lived in the lounge room that was set up as a hospital for him. He was totally dependent for all his personal needs for the last five months of his life.

I concentrated on accepting the nature of the disease, and even with his executive function impairment, I determined that it was his right to make his own decisions. He was terminal, and he had so little choice in anything, that taking what choice he had to force him to use equipment or therapies seemed counterproductive to me if he was to have any quality of life for whatever quantity he had left.

He passed very peacefully, at home with me doing his palliative nursing alone. With the right palliative medications and a calm atmosphere, he had no fear, and we were both at peace.

I have never had any regrets about how I cared for Jim during that 11 months, and now work actively in support of PALS and CALS which gives me a lot of comfort.

Dot's Story

JJ's journey into ALS hell started with back pain. We both pretty much ignored it for the first two or three months. Then he started limping. He went to our GP who ordered an MRI of his lower back. It came back clear and we were happy. The limping only got worse. One day I received a call from the hospital. JJ had fallen at work and landed on his face. They released him after doing a couple of tests and stitching up his head. He was told to stay home for a week and rest up before going back to work. During that time, I noticed that he was tripping more and the limp was getting worse. He was sleeping most of the day. His GP suggested that JJ see a neurologist. The neurologist ordered more tests and sent him to a neuropsychologist. The neuropsychologist found that his motor skills were moderately impaired and his cognition was mildly impaired. She referred him to a neuromuscular physician at Johns Hopkins. They did more testing, including a pulmonary function test. We were in Baltimore for about a week. At the end of the week JJ was told he had ALS. We immediately went to Mayo Clinic for a second opinion and we were told the same thing. We were told to go home and get our financial affairs in order and choose an ALS clinic close to home. JJ never returned to work. We spent the next three months fighting for his long-term disability, getting approved for Social Security and Medicare, and revising our Wills and medical directives. From that point on it has been a blur.

Breathing equipment, walkers, power wheelchair, Hoyer lifts, feeding tube placement, and total exhaustion. We don't have the financial resources to travel because we decided having in-home care a few hours each day was what both JJ and I needed to keep going. As I write this I am constantly glancing at the clock to make sure I check on JJ, blend his food for feeding, set my alarms for hydrating him and get him to the toilet one more time before bed. Both JJ and I will be up several times during the night. Then, tomorrow, we will start all over again with our routine. I'll be able to get out for about two hours tomorrow to grocery shop because a friend will stay with JJ. I'm interviewing home healthcare workers and trying to put together a "team" for JJ but that has been hard. Some of them seem afraid of his equipment and others only want to do certain things and leave the rest for someone else. Some agencies flat out refused to send someone for an ALS patient. So I'm left with finding people willing to be educated and trained by me.

Conclusion

If you are interested in helping people with ALS, there are three very important things you can do. The first is to contact a PALS or CALS to find out how you can assist with their immediate needs and their ongoing needs. In doing this please be realistic on how much time, money, and effort you want to contribute. Once you commit, please honor your promise. Hopefully, this document has some suggestions to get you started. Get as many others involved as you can. This disease is fierce and it is demanding and destructive. If you don't know someone with ALS, contact the local ALS support chapter and ask if there is a PALS who needs something and consider a direct donation to that person.

The second thing you can do is help create awareness of ALS and the burden this disease places on the victims and those around them trying to help. You can do this by giving all your doctors a copy of this document, sending it to your legislators and others with any kind of influence. Don't be afraid to be bold! Send it to newspapers, friends, relatives, and anyone else you think can help.

The third thing you can do is visit the ALS Association at ALS.org and discover other ways to help fight this disease and find a cure.

In doing these things you will become part of the solution for now and part of a CURE for the future.

I would like to acknowledge and thank all the wonderful PALS and CALS from the US, Canada, Australia, Israel, Ireland, and England who were willing to allow me into their lives for the purpose of promoting world-wide awareness of ALS.
----Kim