ALS by MARTHA WILMORE
ALS (Amyotrophic Lateral Sclerosis) or Lou Gehrig's Disease

What is ALS?

It is a progressive neuromuscular disease that eventually destroys motor neurons that connect the brain with the skeletal muscles.

- Skeletal muscles are involved with voluntary movements such as walking and talking.
- The motor neurons transmit the command to move from the brain to the skeletal muscles, which respond by contracting.
- Once a motor neuron degenerates completely the muscle that it controls no longer receives impulses from the brain

Incidence & Prevalence

Approximately 30,000 patients in the United States currently have ALS.

- The disease has no racial, socioeconomic, or ethnic boundaries
- The life expectancy of ALS patients is usually 3 to 5 years after diagnosis.

Symptoms:

Approximately 60% of ALS patients experience muscle weakness and stiffness as the initial symptom. Usually the first muscles affected are those in the arms and legs. Walking or climbing stairs may be difficult.

- Patient may drop things, fall, experience muscle cramps
- May laugh or cry uncontrollably
- Arms and legs may feel especially tired
- If hands are affected, the patient may have difficulty picking up small objects or turning keys.
- Speech problems, such as slurring, hoarseness, or decreased volume may occur
- Swallowing becomes difficult
- advanced patients ….
    - o Experience muscles atrophying, causing spasticity, stiffness, abnormal movements, alterations in gait and manual dexterity
    - o Twitching may occur in the tongue and in affected limbs
    - o Muscle pain & cramps are common
    - o Excessive salivation and difficulty swallowing may cause drooling
    - o Become paralyzed and often require ventilation and surgery to provide a new opening in the stomach (gastrostomy)
    - o When respiratory muscles weaken, the patient may require a ventilator
    - o Loss of respiratory function is ultimately the cause of death in most cases

Medical terminology used:

- Areflexia – absence of spinal reflexes
- Hypotonia – loss of muscle tone
- Fasciculations – muscle twitching
- Atrophy – muscle wasting
- Spasticity – reduced muscle tone or hypotonia & rigidity
- Dysarthria – impaired speech

- Dysphagia – impaired swallowing
- Clonus – rapidly alternating muscle contractions and relaxations

Causes:

- 5 to 10% is suspected to be a genetic defect (DNA defect) … familial ALS
- lack of SOD (superoxide dismutase)
- an autoimmune response where excessive levels of glutamate can overstimulate motor neurons and cause them to die
- Viruses
- Heavy metals
- Enzyme abnormalities
- Neurotoxins (especially in Guamanian ALS)

Helpful, Natural Support:

Diet:

- Eat from the "Healthy Body" diet (at least 50% raw)
- One quart of raw fresh extracted juice is recommended each day
- It is imperative to address allergies
- Avoid caffeine, alfalfa "sprouts", etc. – they are TOO stimulatory
- Avoid "female" foods such as eggs, milk, female chicken, etc. due to higher levels of estrogen levels
- Use non-inflammatory soy protein *Shaklee Energyzing* Protein – at least 6+ tablespoons per day
- *Shaklee Physique for* muscle strengthening
- LOTS of raw juices

Since swallowing can be very difficult, it is helpful to have a G-Tube put into the stomach so food & supplements can be ground and administered through the G-Tube … the earlier this is done the better!

Hormones:

- Have hormone levels checked and help to bring to balance by natural hormone creams applied topically

Detoxify:

- *Liver DTX* is essential to detoxify the liver
- Optiflora- ESSENTIAL
- Ensure proper bowel elimination (use *Herb-Lax*) … with ALS the bowel can become lazy, so help is essential
- Alfalfa is a great detoxifier

Lifestyle:

- Remove all toxic cleaners and personal care products –
  Shaklee products are the only ones that will never injure on human cell!
- Check of leaking amalgams
- Have a hair analysis done to determine if there are is heavy metal toxicity

- Get adequate rest
- Use *Stress Relief* to control stress
- Drink purified water *Get Clean Water* and filter the water in the shower to remove chlorine
- Use digestive enzymes to assist the failing digestive system … *EZ-Gest, Stomach Soothing Complex*

Supplements:

- *NutriFeron* to increase interferon levels
- *Vitalizer Gold*
- *CoQHeart* - CoenzymeQ10 to energize muscle activity
- Antioxidants & Immune Support
    - o *Vitamin E* (has Selenium + Grapeseed Extract)
    - o *Vitamin C*
    - o *Carotomax* – anti oxidant
    - o *FlavoMax* in – anti oxidant
    - o *Immunity Formula* I
- *OmegaGuard, GLA, Lecithin* … use affluently- the essential Oils
- *B Complex*
- *OsteoMatrix* for muscle strengthening and regulation
- *Zinc*
- *Alfalfa* (use affluently)
- *CorEnergy* – an adaptogen for balancing the system

The above was given before *Vivix* or the *Vitalizer* was available. Thus VIVIX is an absolute must for helping the mitochondrial activity to increase in the cells which would help with the improvement of the muscles and slow down or potential stop muscle deterioration.

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Lou Gehrig's Disease - Amyotrophic lateral sclerosis (ALS)

My name is Michele and I want to share with you a little bit about the hope that Chris and I have found. In 1994 (before I knew Chris) Chris was diagnosed with a condition known as Lou Gherig disease, or A.L.S. For those of you who are not familiar with A.L.S., it is a motor neuron disease that causes muscle atrophy as a result of nerve degeneration. In the medical field A.L.S. is known as a terminal illness because the diaphragm muscle which is necessary for breathing atrophies causing shortness of breath and respiratory failure. As a matter of fact, when the neurologist shared the devastating news with Chris he told him, "I'm sorry to tell you this, but it doesn't look good for you." He proceeded to tell him to get prepared and get his will in order. Confused and dazed Chris put on a false mask of courage and walked out of the doctor's office numbly.

There was not much hope offered to him. Some well meaning, but insensitive church friends took Chris to a local cemetery where they priced caskets and toured the burial grounds. They even offered him a funeral planner and advised him to keep his family involved in the funeral arrangements. Others avoided him. His family doctor wouldn't return his calls. It appeared that people expected him to die from this disease. He did too. Why? From his perspective friends and relatives looked on him as a dying individual. Also, the specialist had no course of treatment. In addition, his late grandmother died from A.L.S. and his brother, father, aunt, and four cousins are all currently afflicted with the same illness. As a result, Chris gave in to the disease and waited to die. When my husband Robert and I met Chris in the spring of 1995 he was our son's little league coach. I remember our hearts went out to him when we learned of his condition. "Such a young man," we thought, "with a terminal illness. He has four young sons." You know you never know what kind of curve life can take. A year after we met Chris my husband's heart suddenly stopped one evening as we were getting ready for bed. He died that night at the age of 31 with no previous medical condition. He was in good health supposively.

The autopsy report did not show a cause of death. It was some time after that, I saw Chris at Mc Donald's. I informed him what had gone on in my life and he shared with me the progression of the disease. Soon after, I became his caregiver. I remember seeing the wheelchair and the leg braces in his office. He explained that he needed to use the wheelchair for long distance walking because his legs would get tired and weak at places like the mall or furniture stores. The braces were to help a condition called drop-foot. (The muscle in his leg was weak and his foot would drop down a little causing him to trip and fall over curbs or bumps in the sidewalk.) In addition, the weak muscles in his leg caused cramps< when he would drive too long. As his caregiver I would drive him on errands, help prepare meals, and help him with passive exercises the therapist gave us to help prevent some of the stiffness that goes along with A.L.S. Neither one of us knows exactly when, but somewhere during our friendship grew a very strong love and bond between us. We eventually got married and thus begun our journey of hope and healing.

This new marriage brought a deep sense of joy and happiness for Chris, and with that happiness came a will to live in him so strong that he was not only willing to live, but to actually really work at staying alive longer. Chris remembered taking Shaklee supplements years before his diagnosis and wanted to get started on them again. He looked in the yellow pages and found a distributor. Verna, our upline, had a positive attitude and she encouraged us not to give in to the disease. Since Chris's supplements need was so high, Verna shared with us how to become supervisors so the money we spent on supplements every month would be rebated back in the form of a Shaklee bonus check. Of course, we liked that.

What we enjoyed even more was how good Chris began to feel. We began a program of prayer, special nutrition, exercise, positive mindset acupuncture, and Shaklee supplements**. We began to see an overall improvement in Chris' health. His walking got stronger, the choking and swallowing problems went away, and he does not lose his balance like he used to. We still continue our program. We go on our nightly walks, and now we take long hikes in the Santa Ana Mountains and he is more sure-footed than ever! We are enjoying life and praising God for how good life is, and we are working to improve the lives of everyone who comes in our path.** We want to share the message that no one knows how long we have on this earth except God. I learned that example from Robert's death, no one saw it coming. Then there are those who have been diagnosed with terminal illnesses and go on to live long, healthy lives baffling their doctors.

One thing I have learned is to live each day as if it's our last, as a gift from God. Dedicating each day before Him and asking for His help on how we should live it. He has given us tools, He gives us guidance, he gives us compassion, He gives us hope, and He has given us Shaklee. Let's use those gifts to help others. It is good to encourage people to never give up on a dream or a goal they have. Also, never, ever give in to an illness or seemingly hopeless situation. Envision the positive outcome you are hoping for in your mind. Always have hope. Once you truly have hope inside, you are better able to share that hope with other people who really and need it…..Chris and Michele P

This information is not intended to replace medical care. This information is not intended to diagnose, treator cure.

From ALS Association webpage – FOR YOUR INFORMATION…
WHAT IS ALS (Lou Gehrig's Disease)

The body has many kinds of nerves. There are those involved in the process of thinking, memory, and of detecting sensations (such as hot/cold, sharp/dull), and others for vision, hearing, and other bodily functions. The nerves that die when you have ALS are the motor neurons that provide ***voluntary movements*** *and* ***muscle power****. Examples of voluntary movements are your making the effort to reach for the phone or step off a curb; these actions are controlled by the muscles in the arms and legs.*

The heart and the digestive system are also made of muscle but a different kind, and their movements are not under voluntary control. When your heart beats or a meal is digested, it all happens automatically. Therefore, the heart and digestive system are not involved in ALS. Breathing also may seem to be involuntary. Remember, though, while you cannot stop your heart, you can hold your breath - so be aware that ALS may eventually have an impact on breathing. Although the cause of ALS is not completely understood, the 1990's have brought a wealth of new scientific understanding regarding the physiology of this disease.

Perspective from Hiroshi Mitsumoto, M.D., Cleveland Clinic ALSA Center and Chair of ALSA's Medical Advisory Committee: "In a review of ALS published in the *Archives of Neurology* in 1988, I quote Lewis Thomas. 'The whole field of biomedical science is on the move as never before in the long history of medicine. I don't know what will happen over the next 20 years, but my guess is that we are on the verge of discoveries that will match the best achievement in infectious disease a generation ago.' In ten years - just half of Lewis' prediction - we now know the gene responsible for some familial ALS; we have the first drug we can prescribe for ALS; we have a real animal model for this disease and we have incredibly important knowledge on the cell death mechanisms of motor neurons in ALS. Yes, the progress still appears to be too slow for anyone waiting for a breakthrough, but we are truly on the verge of more exciting discoveries. We have solid reasons for strong hope in ALS." There is no question about **whether** the cause of ALS will be found; it is only a question of **when**.

While there is not a cure or treatment today that halts or reverses ALS, there is one FDA approved drug, Rilutek®, that modestly slows the progression of ALS as well as other drugs in clinical trials that hold promise.

Initial Symptoms of the Disease At the onset of ALS the symptoms may be so slight that they are frequently overlooked. With regard to the appearance of symptoms and the progression of the illness, the course of the disease may include the following:

- muscle weakness in one or more of the following: hands, arms, legs or the muscles of speech, swallowing or breathing
- twitching (fasciculation) and cramping of muscles, especially those in the hands and feet
- impairment of the use of the arms and legs
- "thick speech" and difficulty in projecting the voice
- in more advanced stages, shortness of breath, difficulty in breathing and swallowing

The initial symptoms of ALS can be quite varied in different people. One person may experience tripping over carpet edges, another person may have trouble lifting and a third person's early symptom may be slurred speech. The rate at which ALS progresses can be quite variable from one person to another. Although the mean survival time with ALS is three to five years, many people live five, ten or more years. In a small number of people, ALS is known to remit or halt its progression, though there is no scientific understanding as to how and why this happens. Symptoms can begin in the muscles of speech, swallowing or in the hands, arms, legs or feet. Not all people with ALS experience the same symptoms or the same sequences or patterns of progression. But, progressive muscle weakness and paralysis are universally experienced.

Muscle weakness is a hallmark initial sign in ALS, occurring in approximately 60% of patients. Early symptoms vary with each individual, but usually include tripping, dropping things, abnormal fatigue of the arms and/or legs, slurred speech, muscle cramps and twitches and/or uncontrollable periods of laughing or crying.
The hands and feet may be affected first, causing difficulty in lifting, walking or using the hands for the activities of daily living such as dressing, washing and buttoning clothes. As the weakening and paralysis continue to spread to the muscles of the trunk of the body the disease, eventually affects speech, swallowing, chewing and breathing. When the breathing muscles become affected, ultimately, the patient will need permanent ventilatory support in order to survive.

Since ALS attacks only motor neurons, the sense of sight, touch, hearing, taste and smell are not affected. For many people, muscles of the eyes and bladder are generally not affected. For the vast majority of people, their mind and thoughts are not impaired and remain sharp despite the progressive degenerating condition of the body.

Who Gets ALS
ALS is one of the most devastating disorders that affects the function of nerves and muscles. Based on U.S. population studies, a little over 5,600 people in the U.S. are diagnosed with ALS each year. (That's 15 new cases a day.) It is estimated that as many as 30,000 Americans have the disease at any given time.

Most people who develop ALS are between the ages of 40 and 70, with an average age of 55 at the time of diagnosis. However, cases of the disease do occur in persons in their twenties and thirties. Generally though, ALS occurs in greater percentages as men and women grow older. ALS is 20% more common in men than in women. However with increasing age, the incidence of ALS is more equal between men and women.
Half of all people affected with ALS live at least three or more years after diagnosis. Twenty percent live five years or more; up to ten percent will survive more than ten years.

Forms of ALS
Three classifications of ALS have been described:

- Sporadic - the most common form of ALS in the United States - 90 to 95% of all cases.
- Familial - occurring more than once in a family lineage (genetic dominant inheritance) accounts for a very small number of cases in the United States - 5 to 10% of all cases.
- Guamanian - an extremely high incidence of ALS was observed in Guam and the Trust Territories of the Pacific in the 1950's.

The most common form of ALS in the United States is "sporadic" ALS. It may affect anyone, anywhere.
"Familial" ALS (FALS) means the disease is inherited. Only about 5 to 10% of all ALS patients appear to have genetic or inherited form of ALS. In those families, there is a 50% chance each offspring will inherit the gene mutation and may develop the disease.

Diagnosing ALS

ALS is a very difficult disease to diagnose. To date, there is no one test or procedure to ultimately establish the diagnosis of ALS. It is through a clinical examination and series of diagnostic tests, often ruling out other diseases that mimic ALS, that a diagnosis can be established. A comprehensive diagnostic workup includes most, if not all, of the following procedures: • electrodiagnostic tests including electomyography (EMG) and nerve conduction velocity (NCV)

• blood and urine studies including high resolution serum protein electrophoresis, thyroid and parathyroid hormone levels and 24 hour urine collection for heavy metals

• spinal tap

• x-rays, including magnetic resonance imaging (MRI)

• myelogram of cervical spine

• muscle and/or nerve biopsy

• thorough neurological examination

These tests are done at the discretion of the physician, usually based on the results of other diagnostic tests. There are many diseases that have some of the same symptoms as ALS and many of these conditions are treatable. It is for this reason that The ALS Association recommends that a person diagnosed with ALS seek a second opinion from an ALS "expert" - someone who diagnoses and treats many ALS patients. The ALS Association maintains a list of recognized experts in the field of ALS. See ALSA Centers, ALS Clinics, and the ALS Physician List.

Genetic Testing for ALS

Q. Is ALS hereditary?

A. ALS is directly hereditary in only in a small percentage of families. The majority of patients with ALS (90%) have no family history of ALS, and present as an isolated case. This is called sporadic ALS (SALS), and although there are likely to be genetic risk factors involved, SALS is not directly inherited in a family. Rare exceptions are when familial ALS (FALS) is masked due to an incomplete family history, such as if the patient is adopted. The remaining10% of persons with ALS have a close second family member with ALS, which is referred to as familial ALS (FALS). Currently the best tool to distinguish between SALS and FALS is the family history. A neurologist or genetic counselor will ask whether anyone else has ever been diagnosed with ALS, and if anyone else in the family had progressive walking or speech problems. If so, they will likely ask additional questions to see if the health problems were related to ALS or any number of other causes. They will also inquire about the ages that family members passed away to see if any close relatives passed away at a young age, meaning that a long health history is not available. It is very common to have limited information on one's family, but most families can still be reassured since the majority of instances of ALS are not directly hereditary. Older relatives are often good sources of family history information, and medical records can often be obtained with the help of a hospital's medical release form.

Q. How is FALS inherited?

A. To answer this question, it is helpful to review background information on genetics as it pertains to FALS. Every cell in the human body contains genes. Genes have many functions. Some genes contribute to traits like eye and hair color while other genes are responsible for making proteins that determine how our bodies circulate blood or send nerve signals to muscles. When a gene is disrupted by a change in its sequence, the gene cannot function correctly. Genes are packaged in chromosomes. Chromosomes are present in pairs. The genes that reside within chromosomes are therefore also present in pairs. In each chromosome pair, one chromosome is inherited from the mother and one is inherited from the father. We have 23 pairs of chromosomes, giving us a total number of 46 chromosomes. The first 22 pairs are the numbered chromosomes in which both men and women share them in common. Only the 23rd pair differs between males and women since this pair is the sex chromosome where women typically have two Xs and men have an X and a Y. There are at least 3 different inheritance patterns for FALS. The most common inheritance pattern for FALS is called autosomal dominant. Autosomal means that it is equally likely that a female or male would inherit the gene for FALS because the gene is located on a numbered chromosome that both men and women share in common. Dominant refers to the fact that a person only needs one gene coding for FALS to have a genetic predisposition for ALS. Therefore, a child born to someone who has FALS has a 50% chance to inherit the FALS gene and conversely, a 50%

chance to not inherit the FALS gene. The 1 in 2, or 50% chance, comes from the fact that parents randomly pass on only one member of their gene pair, so that either the gene coding for ALS will be passed on or the gene not coding for ALS will be passed on. Even though parents often feel responsible for their children's health, they have no control over which gene they pass on, just as their parent had no control which gene they passed onto their child. It is also important to remember that inheriting the gene for FALS in no way guarantees that a person will develop symptoms of ALS. If a child does not inherit the gene coding for ALS, they cannot pass it onto their children.

Q. Is there a genetic test for FALS?

A. Yes, although genetic testing is still limited in FALS. Changes in one gene located on chromosome #21 and called superoxide dismutase (SOD1) have been found in about 20% of families with FALS. The SOD1 gene is composed of five regions called exons. If you think of your genetic material as a string of letters that together make up a book of instructions for the human body, the SOD1 gene is one chapter and composed of 5 different pages. SOD1's normal role is to detoxify substances called free radicals, which can be harmful to cells. Changes in the SOD1 gene are thought to create a new but still undefined function that is toxic to motor neurons. Most often, SOD1 changes are inherited in an autosomal dominant fashion. Of particular note is that the majority of families with FALS (80%) will not have a change in their SOD1 gene and therefore, a normal SOD1 genetic test is not informative in a family where an SOD1 change has not been identified. Although researchers are diligently looking for other genes, at this time there is no genetic testing to offer non-SOD1 families. Therefore, the determination that an individual has FALS is typically based on family history rather than a genetic test.

Q. Does a genetic test diagnose ALS?

A. No. Since the vast majority of patients do not have the hereditary type of ALS, diagnosis of ALS is not determined by a genetic test. Instead, a neurologist makes the diagnosis after a review of a person's symptoms, a neurological exam, and results on nerve and muscle function tests. Clinically, FALS and SALS are basically identical.

Q. Who is appropriate for genetic testing?

A. Anyone who has symptoms of ALS in addition to a family history of ALS, such as a parent, grandparent, aunt or uncle, or a brother or sister. Additionally, if one's family history is unknown or a parent passed away at a young age, testing is also appropriate. However, only about 2% of all patients with ALS will have an SOD1 genetic change.

Q. What would the results of the genetic test tell me?

A. A positive test means that the genetic cause of FALS has been identified. Researchers have developed a mouse model with the same genetic change so that they can better understand how a change in the SOD1 gene can lead to the symptoms of ALS. Currently, new therapies are being tried on this animal model to slow or halt the progression of ALS. Although still in the distant future, gene therapy to correct the genetic change is also being researched. A positive test does not change medical treatment at this time and may or may not provide prognostic information. Even though the inheritance may already be established by the family history, an individual may feel furthered burdened by learning they carry a change in their SOD1 gene as concerns for children resurface. Others prefer to have this knowledge and may feel comforted that there is much research aimed specifically at ALS caused by changes in the SOD1 gene. A negative test means only that the genetic cause of ALS has not been identified. However, this does not rule out familial ALS since there are still other unidentified genes that cause ALS in 80% of FALS families.

Q. If I have a family history of FALS, should I have a genetic test even if I don't have symptoms?

A. This situation is called presymptomatic testing. The decision to have presymptomatic genetic testing is highly personalized and often individuals in the same family will disagree whether to pursue it. However, in order for the test to be meaningful, a genetic change in the SOD1 gene needs to first be found in a family member affected with ALS. This will determine whether an unidentified gene is the cause of FALS in the family. When an SOD1 change is not identified in a symptomatic person, presymptomatic genetic testing is not available for other family members. Benefits of presymptomatic genetic testing in ALS is limited by the absence of preventative treatment, the inability to predict the age at which someone who is a gene carrier will get ALS, or even that a gene carrier will definitely get ALS. Since both a negative or positive presymptomatic test result in a known SOD1 family can have a great emotional impact, genetic and psychological counseling is usually required before undergoing such testing. Individuals often consider how learning that they did or did not inherit the predisposing gene would affect their lives, who they would tell about the results, and how relationships may

change depending on the results. Individuals who learn they do not carry the SOD1 change often feel great relief, although they can sometimes wonder why they escaped while another family member did not. They may regret past decisions made based on the presumed at risk status, or find it hard to let go of that part of their identity. Learning that one does carry a predisposing gene, is usually more difficult and that person may need ongoing professional support. Ambiguity is not entirely erased as the question may change from do I carry the gene to when or will I get symptoms? Commitment to friends and family may be strengthened. However, knowledge of the testing by insurance companies or employers is a concern regarding future coverage. A genetic counselor can further discuss the issues involved in presymptomatic testing.

Q. How is the genetic test done?

A. A blood sample is taken and sent to a specialized lab where the genetic material, also called DNA, is removed. Special laboratory techniques allow the SOD1 gene to be replicated and then run on a gel. This generates a series of bands. If a genetic change is present, the bands will be in a different location compared to a control sample, which is known not to have a genetic change in the SOD1 gene. This method is called single strand conformation polymorphism or SSCP for short. Another method called sequencing may be done to confirm the results. Sequencing is able to view the DNA on a finer scale by displaying the actual letters of the "instruction book" so that changes can be seen.

Q. How long does the genetic test take?

A. Because five different parts of the SOD1 gene need to be looked at, the testing usually takes about 2-3 months. The cost is about $300-500 depending on the clinical laboratory that is doing the testing.

Facts You Should Know About ALS

• The onset of ALS is insidious with muscle weakness or stiffness as early symptoms. Inevitable progression of wasting and paralysis of the muscles of the limbs and trunk as well as those that control vital functions such as speech, swallowing and breathing follows. In most cases, mental faculties are not affected. Also, ALS is not contagious. It is estimated that ALS is responsible for nearly two deaths per hundred thousand population annually. More people die every year of ALS than of Huntington's disease or multiple sclerosis.

• A little over 5,000 people in the U.S. are diagnosed with ALS each year. The incidence of ALS (two per 100,000 people) is five times higher than Huntington's disease and about equal to multiple sclerosis. It is estimated that as many as 30,000 Americans may have the disease at any given time.

• The life expectancy of an ALS patient averages about two to five years from the time of diagnosis. Half of all affected live more than three years after diagnosis.

• About twenty percent of people with ALS live five years or more and up to ten percent will survive more than ten years and five percent will live 20 years. There are people in whom ALS has stopped progressing and a small number of people in whom the symptoms of ALS reversed.

• ALS occurs throughout the world with no racial, ethnic or socioeconomic boundaries.

• ALS can strike anyone. Someone you know or love will die from ALS unless a cure or prevention is found.

• Present treatment of ALS is aimed at symptomatic relief, prevention of complications and maintenance of maximum optimal function and optimal quality of life. Most of this, in the later stages, requires nursing management of a patient who is alert but functionally quadriplegic with intact sensory function, bedridden and aware he or she is going to die.

• In 1991 a team of ALSA-funded researchers linked familial ALS to chromosome 21. In 1993 the research team identified a defective SOD1 gene on chromosome 21 as responsible for many cases of familial ALS. Further study indicated over 60 mutations (structural defects) in the SOD (superoxide dismutase) enzyme which alters the enzyme's ability to protect against free radical damage to motor neurons. These studies open possibilities for future therapies or strategies to effectively mediate both familial and sporadic ALS. But much more research on the SOD enzyme is needed. Also, researchers have not ruled out other gene involvement (on other chromosomes) in ALS.

• The financial cost to families of persons with ALS is exceedingly high. It is estimated that in the advanced stages, care can cost an average of $200,000 a year. Patients' and relatives' entire savings are quickly depleted because of the extraordinary cost involved in the care of ALS patients.

• Rilutek®, the first treatment to alter the course of ALS, was approved by the FDA in late 1995. This antiglutamate drug appears to prolong the life of persons with ALS by at least a few months and more recent studies suggest Rilutek® slows the progress of ALS, allowing the patient more time in the higher functioning states. Rilutek is manufactured by Aventis Pharmaceuticals.

Reports from three separate patient databases described long range experience with Rilutek®. All three reports suggest a trend of increasing survival with Rilutek® over time. More studies that are double blind and controlled are needed to confirm these database observations. The trend appears to indicate that longer periods of time than those used in the Rilutek® clinical trials may be needed to see the long-term survival advantage of the drug. An interesting observation was that despite the fact that the Irish government provides Rilutek® free of charge to people in Ireland with ALS, only two-thirds of the patients registered in the Ireland national ALS database reported taking Rilutek®. This paragraph is taken from a report on the 12th International Symposium on ALS/MND.

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From ALS Association webpage