

AMYOTROPHIC LATERAL SCLEROSIS

Amyotrophic Lateral Sclerosis is more commonly known as ALS or Lou Gehrig's disease. ALS is a progressive and fatal disease affecting both men and women. The onset is generally in middle age, and men are one and a half times more likely to suffer from the disease than women. It is estimated that 20,000 to 30,000 people in the United States suffer from ALS, with 5,000 new cases being diagnosed every year (NINDS 2003, ALS Association 2004)

ALS targets the nerve cells located throughout the brain, brain stem, and spinal column, which control voluntary movement. As these nerve cells deteriorate, patients lose control of their muscles, causing the muscles to atrophy and convulse. The muscles affected typically include those of the arms, legs, throat, and eventually lungs. The muscles affected first will vary from patient to patient. Although ALS ravages patients' bodies, it leaves their mental ability unaffected (NINDS 2003).

The prognosis for a patient diagnosed with ALS is bleak. Although at onset the symptoms are generally mild, including cramping and weakness of isolated muscles, the disease progresses quickly. Most patients die from respiratory failure within five years of being diagnosed (NINDS 2003). Due to its debilitating nature, patients with ALS will eventually become completely paralyzed, losing the ability to function on their own, and requiring constant assistance. As their lungs and diaphragm fail, patients will require respiratory assistance. The toll on the patient

and care-giver is enormous. There is no cure for this disease.

The causes of ALS are still unknown. In 5%-10% of new cases, the disease seems to be hereditary. The other 90%-95% of cases have no family history of ALS (MDA Publications 2003). Regarding the hereditary causes of ALS, much genetic research is underway, and a team of researchers recently discovered a gene called ALS4 that causes a type of ALS affecting only juvenile males and women in their thirties. This type of ALS is unique because the patients do not lose the ability to breathe or swallow so they typically live a normal life span (The ALS Society of Canada 2005). Research involving veterans deployed to the Persian Gulf in the 1991 Gulf War may offer new insight about environmental risks and ALS. The research finds that such veterans have developed ALS at nearly twice the rate of veterans not deployed. This research was taken from a study published in *Neurology*, the scientific journal of the American Academy of Neurology (The ALS Association 2004b).

The ALS association estimates that across the United States, 2 deaths per 100,000 can be attributed to ALS (ALS Association 2004). The Agency for Toxic Substances and Disease Registry reported a slightly lower number with approximately 1.71 deaths per 100,000 being attributed, at least in part, to ALS (Noon, Sykes & Hilsdon 2002). The same agency found that between 1989 and 1998 154 people in Bexar County died of ALS. This yields a crude rate of only 1.22 per thousand deaths. Once adjusted

for age of population, the number is close to that of the United States as a whole, at 1.50. One flaw with this study is that the classification system for recording the cause of death for ALS includes both ALS deaths and deaths attributable to other neurological diseases. However, the number of neurological disease deaths not due to ALS is believed to be a relatively small number and to be offset by the number of deaths having to do with ALS where neurological diseases were not named as a cause of death (Noon, Sykes & Hilsdon 2002).

Locally, concern has been expressed about the occurrence of ALS among residents near the former Kelly Air Force Base in San Antonio, and among those who previously worked at the base. In response to these concerns, the Air Force Institute for Environment, Safety, and Occupational Health Risk Analysis conducted a study of 32,000 former Kelly AFB employees. This study concluded that former workers did not have a higher incidence of ALS than other populations living in Bexar County (Ride for Life 2002). The ALSA is pursuing federal money from the Department of Defense to help fund research into possible environmental causes or contributors to ALS (ALS Association 2004c).

Prior to the release of this study, the Agency for Toxic Substances and Disease Registry prepared a paper analyzing possible environmental factors contributing to the number of cases of ALS in other areas of the world. Environmental factors listed in this study include lead and other heavy metals, radiation, dietary exposure to trace elements, agricultural chemicals, and exposure to solvents and other non-agricultural chemicals. While many studies have suggested some relation between these agents and ALS, none have been conclusive. The same report states that, as a whole, Bexar County has an ALS rate similar to the state of Texas and the United States.

In San Antonio, the ALS Association-South Texas Chapter works to promote research and help those suffering from ALS. The University of Texas Health Science Center in San Antonio

houses a clinic specializing in the care of patients with ALS. The cost of treating ALS varies as widely as the symptoms. According to a recent study, the costs during the initial phase of diagnoses generally range from \$10,000-\$20,000, as doctors rule out other diseases. As the disease progresses, the financial burden on the family will depend on how much care a patient needs. Some patients are cared for by a family member, causing the family to forgo the income of both the patient and the caregiver. Other patients have to enter full time care at a hospice or nursing facility that may or may not be covered by insurance or Medicare. Estimates of the cost of treatment in the final stages range from \$80,000-\$200,000 per year (Barisano and Kramer 1998).

Although research is being conducted on many drugs, to date, the FDA has approved only one drug, riluzole (or Rilutek), to fight the progression of ALS. This drug costs about \$700 a month, and unfortunately only “has a modest effect in prolonging survival” (MDA Publications 2003). Along with other preliminary drugs, physicians also recommend a number of over-the-counter substances, including vitamin E, coenzyme Q10, and creatine. These substances have shown promising improvements in ALS, and further studies are continuing (MDA Publications 2003b).

However, there are many approaches to maintaining a higher quality of life for those diagnosed with ALS. The Muscular Dystrophy Association (MDA) and the ALS Association support centers across the United States for the treatment of patients suffering from diseases such as ALS (MDA Publications 2003). In these centers, teams of doctors, speech therapists, occupational therapists, physical therapists, and pharmacists work together to keep the patient as mobile and comfortable as possible. For example, occupational therapists can help modify patients' homes with ramps and other amenities to help them maintain some independence as their condition worsens. Physical therapists help those suffering from ALS to strengthen the muscles they can still

control, without overexerting them. Stretching and flexibility exercises help decrease the twitching and muscle contractions that generally accompany the paralysis and atrophy of muscles (NINDS 2003). Speech therapists work with the patient to maintain clear and audible speech while vocal muscles deteriorate. Teaching patients to use speech synthesizing equipment before they lose the ability to speak, helps eliminate some of the stress induced by the ultimate loss of speech. Furthermore, respiratory therapists help assist patients who experience a decrease in breathing ability. A respiratory therapist is needed to measure on a regular basis the breathing capacity to provide appropriate care. When the breathing capacity drops below 50%, one should consider a permanent hook-up to a ventilator (Life Extension 2004).

Studies have shown promising results for using stem cells to produce nerve cells to counter those destroyed by ALS. In the study, stem cells were transplanted into the damaged area of the spinal columns in rats with spinal injuries. The stem cells yielded nerve cells that integrated into the spinal cord. Rats that received the transplants were able to perform better on tests of movement and strength than those who did not, showing the new nerve cells had improved their condition. While this experiment was conducted on rats with spinal cord injuries, scientists believe the same technology could benefit humans who have lost nerve cells to ALS (Society for Neuroscience 2003).

Studies are also underway to explore the benefits of the supplement creatine and the antibiotic minocycline in the fight against ALS. One experiment found that using creatine and minocycline separately increased survival in mice by 12% and 13% respectively, but when combined the survival rate increased even more, to 25%. The effects of these promising substances on humans are under investigation now (NINDS 2003b, Ride For Life 2003).

Like many disorders involving the brain and spinal column, research is being conducted in

hopes of discovering the cause of and cure for ALS. Until a cure is found, those suffering from ALS must do their best to mitigate the effects of the disease and lessen its complications.

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