

From the Desk of  
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# Amyotrophic Lateral Sclerosis (ALS) and End of Life Care



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## Introduction

Amyotrophic Lateral Sclerosis (ALS) is commonly known as Lou Gehrig's disease, named after the famous first baseman who played for the New York Yankees between 1923 and 1939. ALS is a devastating terminal neurodegenerative disease that has a highly predictable clinical course. It is also known as a motor neuron disease and is a progressive, incurable, neuromuscular disease that attacks nerve cells and pathways in both the brain and spinal cord.

Motor neurons reach from the brain to the spinal cord and then from the spinal cord to a muscle or gland. When motor neurons die, the brain can no longer start or control muscle movement. People with ALS gradually lose the ability to move and will eventually be overtaken by complete paralysis. Towards the end of the disease, muscles used to breathe and swallow will also become affected.

## Who is at Risk for Developing ALS?

Roughly 20,000 Americans have ALS and 6,000 new cases are diagnosed each year. Symptoms begin between the ages of 40 to 70, but there are cases that develop before and after these ages. Most people will unfortunately survive only 3 to 5 years after the diagnosis is confirmed and 10% will live 10 or more years with the disease. ALS appears to be more prevalent in men. Between 5% and 10% of cases are inherited and are known as "Familial ALS". The other 90% to 95% are known as "Sporadic ALS", and there is no known cause.

## Symptoms

People usually become first aware of the onset of ALS when their hands become clumsy, which then causes difficulty performing minor tasks such as handwriting or unlocking a door with a key. Other people may experience weakness of the legs which can lead to tripping, stumbling or falling. ALS is usually present for some time before any signs or symptoms are recognizable. Generalized fatigue may also be an early sign. When symptoms start to occur, they begin in the hands and feet and then travel towards the center of the body. One side is initially more affected than the other side.

Some symptoms that may occur as ALS progresses include:

- Stiffness
- Occasional jerking of arms or legs
- Twitching
- Pseudobulbar affect (uncontrollable laughter or crying)
- Depression
- Drooling
- Cognitive impairment

- Changes in behavior
- Frontotemporal dementia (FTD)
- Deterioration of personality and language skills

## Medications

Two medications have been proven helpful in slowing the progression of ALS and extending the life of those who have been diagnosed with the disease. Edaravone (Radicava) is administered through an IV and is a powerful antioxidant that can prevent damage to nerve cells caused by free radicals. It is currently not known how it works to slow the progression and has bruising, unsteady gait and headache as common side effects.



Riluzole (Rilutek) is taken orally and help reduce damage to motor nerves by reducing the amount of glutamate in the body. Glutamate carries chemical messages to the nerves but when there is too much, it can damage cells. The most common side effects of Riluzole are gastrointestinal distress, dizziness and bruising.

## Special Care Issues during Late Stage ALS

There are numerous and sometimes complex concerns when providing care to persons with ALS. Vitamin A (Retinol) is to be avoided because expression of retinoid signaling genes is altered in ALS and this may contribute to motor neuron loss. On the other hand, Vitamin B12 and Folate are thought to reduce the level of homocysteine, which is involved in the formation of free radicals, excitotoxic amino acid-mediated damage and cytosolic calcium accumulation. Homocysteine is believed to be a biomarker of ALS.

Persons with ALS have lower Vitamin D and may benefit from 2000 IU per day. Vitamin E has been shown in pre-clinical studies to delay onset of ALS but does not affect survival. L-Carnitine has been shown in studies to delay onset of symptoms, deterioration of motor activity and extends the life expectancy.

Weight and diet are two of the most important aspects of caring for people with ALS. Those having a higher BMI may survive longer than those with a lower BMI. Maintaining a healthy weight can be a challenge due to swallowing difficulties, bulbar movement and paralysis. Most times, liquid nutritional supplements are recommended. Cannabis is a powerful antioxidant, anti-inflammatory and neuroprotectant and is beginning to play an important role in the care of people living with ALS. Mild to moderate resistance exercise may be beneficial to people living with ALS.

Spasticity is a common care problem that requires treatment. Hypersialorrhea or hyper-salivation and excessive drooling, is an embarrassing and frustrating condition that will also need to be addressed. Communication will deteriorate and can be compensated with the use of augmentative or alternative communication devices (AAC).

Pulmonary issues will also require frequent intervention as respiratory failure is the leading cause of death in ALS. Other special care issues include pain management, immobility and quality of life.

## The Beginning of End of Life Care

ALS is a very difficult disease for everyone involved including the person diagnosed with it, family and friends, and caregivers who provide care on a daily basis. There is a point where palliative care transitions to the need for hospice care. The person's self-determination and autonomy are top priorities, and a person-centered approach is a must. Hospice care will address every need including physical, emotional, social, and spiritual. Care will be coordinated by physicians, nurses, social workers, chaplains, volunteers, and other hospice professionals. Many needs of the individual and family will be addressed including education, bereavement, decision-making, symptom management and communication. Hospice services can make a significant difference at the end of an individual's life.

## Final Words

Amyotrophic Lateral Sclerosis or ALS, is an overwhelming and highly distressing neurodegenerative disease, for which there is no cure. It begins with clumsy hands and trembling legs and ends in complete paralysis, literally trapping the person within his or her own body. The ability to walk, talk, breathe or even blink an eye will eventually become lost. ALS is a particularly cruel disease and everyone surrounding the individual suffers. Most people will live with the disease for 3 to 5 years and some will live up to 10 years. Treatment may help for a while, but when it no longer does, hospice will be there to provide care at the most crucial time in the person's life.