Amyotrophic Lateral Sclerosis
Motor Neuron Disease
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Lou Gehrig's Disease

What is the Disease?

ALS is an extremely deadly disease affecting the nerve cells that control the victim's voluntary muscles. These nerves shrink and eventually die, leaving the muscles without stimulation. As these muscles go without stimulation, they too eventually shrink and die. The victim progressively weakens to the point of complete paralysis of all voluntary muscles and some involuntary muscles, such as breathing and swallowing, and soon after this point, death is inevitable.

'A' means "Without"
'Myo' means "Muscle"
'Trophic' means "Nourishment"
'Lateral'
refers to uneven development of symptoms between right and left sides
'Sclerosis'
refers to "destruction" of tissue

The History of ALS

A French doctor named Charcot first identified ALS in 1874. It is one of the most devastating diagnoses a person can receive. ALS is said to start between the years of 40 and 70, with the exact average being 45.6 years old.
The most classic case of Amyotrophic Lateral Sclerosis is Lou Gehrig. Lou Gehrig was a New York Yankees first baseman, who from 1923 to 1939, had never missed a game and had a life time batting average of .340. However, the symptoms of ALS emerged in 1938, and in 1939, he was diagnosed with the disease. At that time doctors knew little to nothing about the disease and the only suggested treatment was the untested vitamin E. So Gehrig ate a daily plate full of garden grass, until June 2, 1941 when he died at the age of 37.
ALS affects approximately 1 out of every 100,000 people. In the United States there are around 30,000 Americans affected by ALS, and 3,000 more are diagnosed with the disease each year, with men being affected slightly more than women, and in some cases, running in families. However while this is the same number of new cases as Multiple Sclerosis, Multiple Sclerosis affects around 350,000 Americans. The difference is that 50% of ALS patient's die within three years, and 80% die within five. The disease is in some ways quite similar to Alzheimer's except with Alzheimer's you have a body walking around with a diseased brain, whereas with ALS you have a healthy brain trapped inside a diseased body.

Symptoms

About one-third of those with ALS become aware of their disease when their hands become clumsy, causing difficulty performing anything needing fine finger movements. Another third find a weakness in their legs and may trip because of a mild foot drop. The remaining one-third notice slurring in their speech or difficulty swallowing. Because all of these symptoms happen naturally, it is generally not characterized as ALS until the symptom progressively worsens. This happens as the affected area's muscle cells deteriorate, resulting in muscle tenseness. Frequently one side of the body is affected first and it then gradually passes to the other side. Muscles in the eyes, anus and bladder are generally left

unaffected.

Diagnoses

As there is no known way to prevent this disease, there is also no specific clinical test to identify ALS. It generally involves a physical examination, perusing through the patient's medical history, and neurological testing. To test muscle activity specialists often use an EMG, or electromyogram, and will often use CT scans, MRIs, and thorough blood examination. There is also a recently developed SOD1 scan, the gene now thought to be the cause for ALS, especially familial ALS. Only 20%, however, of patients with familial ALS show positive on the SOD1 scan.

Progress of ALS

Until very recently very little was known about ALS, either what started it or how to treat it. Currently there are 3 types of ALS: classic (sporadic), familial, and the Mariana Island.

Classic ALS accounts for 90-95% of ALS patients in the U.S. The infrequent familial form (FALS) is inherited and if your parents had FALS there is a 50/50 chance you will have it as well. The Mariana Island form is a rare form of ALS found in patients taken from Guam and Japan. ALS appears evenly across the globe except in the Mariana Islands in the West Pacific and the Kii Peninsula of Japan where it is unusually high.

Back during Gehrig's time little else besides vitamin E was even considered a "potential" therapy, and there were only guesses as to the cause of the disease until 1991 when evidence linked FALS to chromosome 21. Then in 1993 the same research team identified a defective SOD1 gene on chromosome 21 as being responsible. It is now known that structural defects in the Super Oxide Dismutase, or SOD, enzyme reduces the ability to protect against damage to motor neurons.

Treatment

Traditionally doctors were unable to subscribe anything other than a good source of Vitamin E, exercise and a healthy mind. However, in June of 1996 the Food and Drug Administration passed the first drug for ALS. The drug Riluzole was successful in lengthening the life-time of ALS patients, especially those with FALS. However, there is still no way to dampen the symptoms or prevent those who don't have it, from getting it. This still was a big step for ALS and there are now 21 countries that have approved Riluzole, including the Czech Republic and all 15 members of the European Union. Gabapentin is also similar to Riluzole and is being tested for approval by the FDA. More importantly, a drug known as Myotrophin is being tested as well by the FDA and may be the first drug to slow the progress of paralysis. Because Myotrophin acts differently than Riluzole, they, hopefully, can be used in synch as well.

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