Mental Health Association of Northern Kentucky

A LEADER IN PROVIDING ADVOCACY, EDUCATION, AND SERVICES THAT PROMOTE MENTAL WELLNESS

EDUCATION AND SCREENING OUTREACH PARTNER FOR THE NATIONAL INSTITUTE OF MENTAL HEALTH AND NATIONAL MENTAL HEALTH ASSOCIATION

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AMYOTROPHIC LATERAL SCLEROSIS

What Is ALS?

Amyotrophic lateral sclerosis (ALS), often referred to as 'Lou Gehrig's disease<" is a progressive fatal neuromuscular disease that attacks nerve cells and pathways in the brain and spinal cord. Motor neurons, among the largest of all nerve cells, reach from the grain to the spinal cord and from the spinal cord to muscles throughout the body with connections to the brain. When they die, as with ALS, the ability of the brain to start and control muscle movement dies with them. With all voluntary muscle action affected, patients in the later stages are totally paralyzed. In most cases, mental faculties are not affected. A-myo-trophic comes from the Greek language. "A" means no or negative. "Myo" refers to muscle, and "trophic" means nourishment.—"No muscle nourishment." When a muscle has no nourishment, it "atrophies" or wastes away. "Lateral" identifies the areas in a person's spinal cord where portions of the nerve cells that nourish the muscles are located. As this area degenerates, it leads to scarring or hardening ("sclerosis") in the region.

Incidence of ALS

ALS is one of the most devastating of disorders, which affect the function of nerves and muscles. The incidence of ALS is about 2 per l00,000 population. Thus, based on the 2000 U. S. Census, some 5,600 people in the U.S. are newly diagnosed with ALS each year. (That's about 15 new cases a day.) It is estimated that as many as 30,000 Americans may have the disease at any given time. Most who develop ALS are between the ages of 40 and 70. There have, however, been many cases of the disease attacking persons in their twenties and thirties. Generally, though, ALS occurs in greater percentages as men and women grow older. It was once believed that men developed ALS more frequently than women. That no longer appears to be the case, and today both sexes are affected in nearly equal numbers. With recent advances in research and improved medical care, many ALS patients are living longer and more productive lives. Half of all affected live at least three years or more after the diagnosis. Twenty percent live five years or more; up to ten percent will survive more than ten years.

Forms of ALS

The most common form of ALS in the United States is known as "sporadic" ALS. It may affect anyone, anywhere. "Familial" ALS suggests the disease is inherited, although not heredity pattern is known to exist in the majority of ALS cases. Only about five to ten percent of all ALS patients appear to have some king of genetic or inherited component. In those families, there is a 50 percent chance the offspring will have the disease. Other terms which have been used to categorize variants of the classical form of ALS include spinal muscular atrophy, progressive bulbar palsy and lateral sclerosis. Other variants of ALS whose prognosis is better and whose relationship to ALS is not yet determined include primary lateral sclerosis, juvenile muscular atrophy and benign facial amyotrophy.

Symptoms of ALS

Early symptoms vary with each individuals, but usually include tripping, dropping things, abnormal fatigue of the arms and/or legs, slurred speech, muscle cramps and twitches and uncontrollable periods of

laughing or crying. The hands and feet may be affects first, causing difficulty in 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 of the body, the disease eventually affects swallowing, chewing and breathing. When the breathing muscles are attacked, the patient faces permanent ventilatory support in order to survive. Since ALS attacks only motor neurons, the senses of sight, touch, hearing, taste and smell and muscles of the eyes and bladder are generally not affected. In most cases the mind is not impaired and remains sharp despite the progressive degenerating conditions of the body.

Diagnosis of ALS

ALS is a very difficult disease to diagnose. There is no one test or procedure to ultimately establish the diagnosis of ALS. It is through a clinical examination and series of diagnostic test, often ruling out other diseases that mimic ALS that a diagnosis can be established. A comprehensive diagnostic workup include most, if not all, of the following procedures: electrodiagnosic tests including electromyography (EMG) and nerve conduction velocity (NCV), blood and urine studies including high resolution serum protein electrophoresis, thyroid and parathyroid hormone levels, 24 hour urine collections for heavy metals, spinal tap and x-rays, including magnetic resonance imaging (MRI) and/or myelogram of cervical spine, and muscle and/or nerve biopsy. 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; some of them are treatable. It is for this reasons that The ALS Association recommends that a person diagnosed with ALS seek a second opinion from an ALLS "expert," someone who sees many ALS patients. The Association maintains a list of recognized experts in the field of ALS. This is not meant to imply that other neurologist cannot make a diagnosis, only that physicians referred by ALSA see many ALS patients.

The Search for Answers

The cause, cure or means of prevention of ALS are presently unknown. The disease was first described in detail in 1869 by the noted French neurologist Jean-Martin Charcot. Subsequent research has been unable to pinpoint the cause of ALS, but a number of hypotheses have been advanced. Some of these have dealt with infections causes, the autoimmune function, heredity, toxic substances, chemical imbalances in the b body and nutrition. More recent scientific studies suggest excitotoxic injury of motor neurons, free-radical-medicated oxidative injury to nerve cells, inflammation and immune response, premature programmed cell death (perhaps as a result of the first three items), environmental factors and "risk" or susceptibility genes Many investigators now believe that the answer to ALS will be multi-faceted.

Treatment Today

Although there is not yet a cure for ALS, much can be done not only to help patients live with the disease, but live more productively and independently. The concept of ALS as a hopeless disease is fast giving way to an approach that emphasizes the treatment of a patients symptoms. This can improve the quality of life for the patient and help him or her develop a positive attitude about being part of the management care team. Physical therapy, rehabilitation techniques, and assistive devices are helping patients learn how to work around the weakness and functional disability caused by the disease. In addition, studies have revealed that some compounds may alter the progression or course of ALS. Rilutek was approved in 1995 as the first drug to alter the course of the disease.

There is information available today. There is direction. There is hope.

What is The ALS Association?

The ALS Association is the *only* national not-for-profit voluntary health organization dedicated *solely* to the fight against *amyotrophic* lateral sclerosis. Its mission: to find a cure for an improve living with ALS. From its

beginnings in 1972, the organization has grown extensively throughout the country. Through its National Office, and growing network of local volunteer chapters and support groups, The ALS Association wages its battles against the disease on four important fronts:

--Encouraging, identifying, funding and monitoring cutting-edge research worldwide into the cause, means of prevention and possible cure of ALS

--Assisting ALS patients and families through referrals for counseling, training and support of how to cope with this devastating disease; providing clinical care regardless of face, color, creed or financial status through its nationwide network of ALSA Centers.

--Serving as the national information resource on ALS for the medical profession, patients and family members.

--Educating the public as to the gravity of this problem to stimulate public support in the search for a cure.