



X-Plain *Amyotrophic Lateral Sclerosis* **Reference Summary**

Introduction

Amyotrophic Lateral Sclerosis, or ALS, is a serious neurological disease that affects the ability to move. It is also called Lou Gehrig's disease.

About 20,000 Americans have ALS, and 5,000 more are diagnosed with it each year. ALS affects people of all races and ethnic backgrounds. It mostly affects people between the ages of 40 and 60 years old. Men are more likely to get ALS than women.

This reference summary will help you understand ALS and its treatment options.

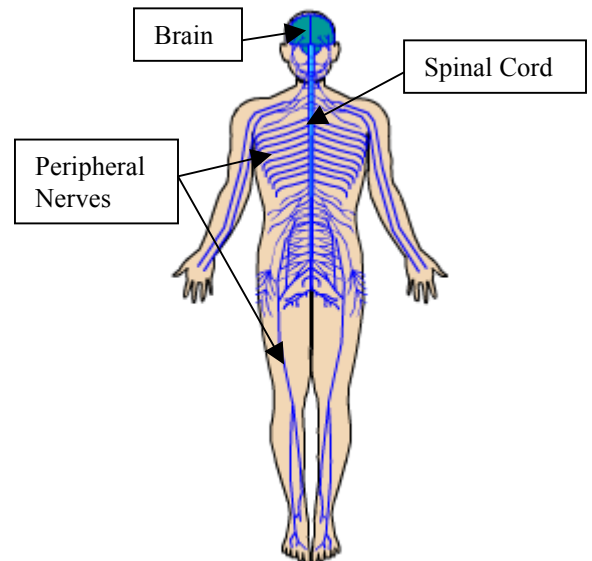
Anatomy

The brain and spinal cord are called the central nervous system.

The nerves in the body, such as the arms, legs, chest, abdomen, and pelvis, make up the peripheral nervous system. The brain is similar to a very complex computer that processes input from our senses and then tells the body how to respond, by talking or moving.

The main cells that make up the brain and the spinal cord are called neurons. The neurons are the thinking cells of the brain. They communicate with each other by sending electric signals through wires called axons.

Neurons that control our muscles are called motor neurons. The axons of motor neurons cause our muscles to contract, which helps us adapt to our environment by talking, walking, chewing, and running.



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ALS

ALS attacks the neurons that control muscles, the motor neurons.

Messages from motor neurons in the brain called upper motor neurons are transmitted to motor neurons in the spinal cord called lower motor neurons, then passed on to the muscles.

With ALS, both the upper motor neurons and the lower motor neurons die and stop sending messages to muscles.

Symptoms

Symptoms of ALS usually occur very gradually. At first, they may be mistaken for symptoms of other less serious diseases.

Symptoms of ALS are caused by destroyed motor neurons. The muscles affected by the destroyed motor neurons gradually weaken, waste away, and twitch.

Early symptoms of ALS affect the parts of the body where the motor neurons got damaged first.

In some cases, ALS affects only one leg at first. Patients notice that they are awkward when they walk or run or that they stumble more often.

Some ALS patients notice the first signs of ALS in their hand or arm. They may find that simple tasks such as buttoning a shirt, writing, or turning a key in a lock are difficult. Other patients notice speech problems.

ALS causes a wide range of disabilities. Eventually, the brain's ability to control voluntary movement is lost. Patients lose strength and the ability to move their arms, legs, and body.

When muscles in the diaphragm and chest wall no longer work, ALS patients cannot breathe without support from a machine.

Most people with ALS experience major respiratory problems within 3-5 years of the first symptoms.



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Since ALS only affects motor neurons, the disease does not impair personality, intelligence, or memory.

ALS does not affect the ability to see, smell, taste, hear, or recognize touch. ALS patients usually retain control of eye muscles and bladder and bowel functions.

Causes

The cause of ALS is unknown. In 90-95% of all cases, it occurs randomly.

Most ALS patients do not have a family history of it, nor do family members have an increased risk for developing ALS. About 5-10% of all ALS cases seem to be genetic.

Researchers have been able to identify the gene related to some cases of familial ALS and are trying to develop a treatment.

Some scientists believe that ALS is caused when the body's immune system attacks the neurons because it has mistaken them for foreign cells.

Diagnosis

A neurologist usually diagnoses ALS after reviewing a very detailed medical history and doing a physical examination.

ALS is usually difficult to diagnose early because it can appear to be another disease, such as spinal cord tumors, multiple sclerosis, or nerve compression.

As the symptoms of ALS progress, it becomes easier to diagnose.

ALS can be diagnosed using electric tests on the muscles and nerves. These tests are called EMG and NCV, for electromyogram and nerve conduction velocity.

Studies of spinal fluid obtained by performing a spinal tap can rule out other disease and can point to ALS.

MRIs and CAT scans help rule out diseases other than ALS that could be causing symptoms.

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Treatment

There is no cure for ALS. However, the Food and Drug Administration, or the FDA, has approved the first drug treatment for ALS called riluzole.

Scientists believe that riluzole reduces damage to motor neurons. It also prolongs life by several months, mostly in patients who have trouble swallowing.

Riluzole does not reverse the damage already done to motor neurons.

Other treatments for ALS relieve symptoms and improve the patient's quality of life. To provide these other treatments, health care professionals team up, including doctors, pharmacists, therapists, social workers, and home care specialists.

Working with patients, healthcare teams can design an individual plan of medical and physical therapies. They can also provide equipment to keep patients as mobile and comfortable as possible.

Doctors can prescribe medications to reduce

- fatigue
- muscle cramps
- excess saliva
- phlegm

Drugs are available to help ALS patients with pain, depression, sleep problems, and constipation.

ALS patients who have difficulty speaking may benefit from working with a speech therapist.

Physical therapy and special equipment or devices such as ramps, braces, walkers, and wheelchairs can help ALS patients keep their independence and remain mobile.

When patients can no longer get enough nourishment from eating, a feeding tube may be inserted into the stomach. The use of a feeding tube reduces the risk of choking.



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A feeding tube also reduces the risk of pneumonia that could result from inhaling food and drinks into the lungs. A feeding tube is not painful and does not prevent patients from eating food by mouth.

When breathing muscles become weak, a breathing machine may be used at night to help the patient breathe while sleeping.

Eventually, a machine may be needed to inflate and deflate the lungs all the time. This machine is called a respirator.

Social workers, hospice nurses, and home care specialists help with the medical, emotional, and financial challenges of coping with ALS, especially during the final stages.

Summary

Amyotrophic lateral sclerosis is a very serious neurological disease that affects the body's ability to control voluntary muscles.

There are no cures for ALS. However, research and new forms of treatment are being developed.

Knowing about ALS and its progression will help patients make informed decisions concerning their health care.



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