Signs and Symptoms

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ALS affects the upper motor neurons, which are in the brain, and the lower motor neurons, which are in the spinal cord and brainstem. Upper motor neuron degeneration generally causesspasticity (tightness in a muscle), while lower motor neuron degeneration causes muscle weakness, muscle atrophy(shrinkage of muscles) and twitching. These can occur in combination in ALS, as upper and lower motor neurons are being lost at the same time.

ALS can affect people of any age, though it usually strikes in late middle age. ALS usually announces itself with persistent weakness or spasticity in an arm or leg, causing difficulty using the affected limb. Sometimes the problem originates in the muscles controlling speech or swallowing. It isn’t unusual for people to ignore such problems for some time at this stage, or to consult a physician who may be relatively unconcerned.

However, the disease — if it’s truly ALS — generally spreads from one part of the body to another (almost always in parts adjacent to each other) so that eventually the problem can no longer be ignored.

It’s at this point that people usually are referred to a neurologist, who will consider ALS among many other possible diagnoses.

The involuntary muscles (such as those that control the heartbeat, gastrointestinal tract, bowel and bladder function) and sexual functions are not directly affected in ALS. However, prolonged inability to move and other effects of ALS can have an indirect impact. Hearing, vision and touch generally remain normal.

Nor is pain a direct consequence of ALS, although pain can occur as a result of immobility and its various complications, especially if precautions such as daily range-of-motion exercises are not undertaken.

Fasciculations are a common symptom of ALS. These persistent muscle twitches are generally not painful, but can be annoying or interfere with sleep. They are the result of the ongoing disruption of signals from the nerves to the muscles that occurs in ALS.

Some with ALS experience painful muscle cramps, which can sometimes be alleviated with medication.

Some people with ALS undergo alterations in their thinking or may exhibit uncharacteristic behavior changes, often referred to as frontotemporal dementia, or FTD. However memory loss, a hallmark of Alzheimer's-type dementia, is generally not a feature of most cognitive changes in ALS. Instead, the person with ALS might be irritable, inconsiderate, apathetic, ritualistic, impulsive or otherwise act in uncharacteristic ways.

Another potential ALS symptom — not experienced by all — is a temporary lapse of control over emotional expressions such as laughing or crying, a phenomenon called pseudobulbar affect. Laughing or crying bouts, often triggered by the smallest of things, are more related to the disease process rather than to actual feelings of happiness or sadness. Medications such as Nuedexta and various other strategies can help manage this symptom.

For more about the progression of ALS symptoms over the full course of the disease, see