

Types of ALS

An estimated 90% or more of all ALS cases are considered sporadic, which means there is no known cause or family history of ALS. Up to 10% of all ALS cases are familial, which means other family members are affected and it is inherited from a parent.

Beyond these two categories, there are subtypes, or forms, of motor neuron diseases that are classified by which part of the motor neuron system is affected.

For traditional ALS, both the upper and lower motor neurons are affected. For primary lateral sclerosis (PLS), just the upper motor neurons are affected. For progressive muscular atrophy (PMA), just the lower motor neurons are affected. Pseudobulbar palsy (PBP) is a condition that affects the motor neurons that control speaking, swallowing, and chewing.

Primary Lateral Sclerosis (PLS)

PLS is a rare motor neuron disease that only affects the upper motor neurons. PLS tends to progress more slowly than ALS and has a longer life expectancy. Because the initial symptoms of PLS and ALS can be similar, an initial PLS diagnosis can change to an ALS diagnosis if lower motor neuron involvement becomes apparent within 2-4 years. Progression of PLS varies from person to person. Symptoms can include leg and arm weakness, muscle stiffness (spasticity), problems with balance, and difficulty speaking and swallowing.

Progressive Muscular Atrophy (PMA)

PMA is a rare motor neuron disease that affects the lower motor neurons and has similarities to ALS. Symptoms vary from person to person, but common symptoms include twitching (fasciculations), muscle weakness, and atrophy. PMA usually progresses more slowly than traditional ALS and more quickly than PLS. An initial PMA diagnosis can change to an ALS diagnosis if symptoms associated with upper motor neuron damage develop.

Pseudobulbar Palsy (PBP)

This condition starts by affecting the upper motor neurons that control speaking, swallowing, and chewing. It is also associated with sudden episodes of involuntary laughing or crying. People with pseudobulbar palsy often, but not always, go on to develop ALS. ALS that starts in the bulbar region can progress differently than ALS that begins in the limbs and usually requires earlier interventions for challenges associated with speaking, swallowing, and eating.

What are motor neurons?

Motor neurons are nerve cells in the brain that send messages to voluntary muscles throughout the body. When ALS causes motor neurons to deteriorate, the messages are disrupted, which leads to muscle weakness, atrophy, and ultimately paralysis.

Upper motor neurons are located in the upper prefrontal cortex of the brain, and lower motor neurons are located in the brainstem and spinal cord. Lower motor neurons do not control the lower body and upper motor neurons do not control the upper body.

In order to control movement, the upper motor neurons carry the message from the cortex down through the lower brain and spinal cord where they contact the lower motor neurons that supply nerves to the muscles that are to be moved. The lower motor neuron then transmits the information to the muscle to move.

Your neurologist—ideally one who specializes in ALS and works at an ALS clinic—should be able to answer any questions you have about the type of ALS you may have and further explain your diagnosis and disease progression.

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