The short version: There are several motor neuron diseases "MNDs"--(diseases that directly damage the nerves that control your muscles). PLS (primary lateral sclerosis) and ALS (amyotrophic lateral sclerosis) are two of them. PLS is less common and less severe than ALS.

The most common reason for being told you have PLS rather than ALS is that your legs, where PLS usually begins, are stiff and spastic (twitchy/crampy) rather than weak and wasted, and because your EMG didn't show damage to the lower motor neurons as ALS does. You were probably told your EMG was "clean" or had minimal concerns.

If you are told you have PLS, it will take years to know if you really have that, or ALS.

If your disease begins as "PLS," even if you later progress to "ALS," you are less likely than other ALS patients to have serious breathing problems. Therefore, PLS is not generally considered fatal, though weakened muscles make falls a risk.

PLS vs. ALS

Neurons are nerve cells. Nerves send electrical signals to muscles to tell them to move, and also send signals to the skin and other organs in a way that causes us to feel. The sensory neurons that cause us to feel pain, heat, tingling are not usually affected by PLS or ALS, which are both *motor* neuron diseases.

But there are major differences between ALS and PLS. In PLS, only upper motor neurons [that communicate with the lower motor neurons that in turn direct muscle movement) are affected. Thus, much of what happens in PLS is "resistance to movement," causing stiffness, spasticity and balance issues.

In ALS, both lower and upper motor neurons are involved, though not necessarily equally. The lower motor neuron damage that ALS causes means more weakness, atrophy and frequently breathing problems than PLS.

In PLS, voluntary muscles (muscles that you can decide to move) are affected, which may include the muscles that control urination, whereas in ALS, *involuntary* muscles are affected, too. So, in addition to the muscles affected by PLS, ALS often affects the muscles that *initiate* breathing, which is why breathing problems in ALS are more severe. Also, unlike PLS, ALS can affect the muscles that let the eyes blink or close, and may also affect the muscles that control bowel movements.

In addition to slower progression than ALS, PLS symptoms can be worse under certain conditions such as stress, exertion and anger (ALS is fairly constant from hour to hour).

PLS signs and symptoms

Signs and symptoms of primary lateral sclerosis (PLS) usually take years to progress. Usually, it starts with stiffness, weakness and spasticity (spasms) in your legs. Sometimes but less often, PLS begins in your tongue or hands and then progresses down your body to your legs. Other symptoms typically follow:

- Weakness and stiffness usually progressing to your trunk, then your arms, hands, tongue and jaw
- Tripping, difficulty with balance and clumsiness

- Hoarseness, problems with speaking, slurred speech and drooling caused by weak facial muscles. Complete loss of speech is rare.
- Difficulties with swallowing and breathing, usually later in the disease
- Like ALS, emotional lability where you veer between laughter and tears (EL) is possible.
- Cognitive impairment (thinking, decision and memory problems) can occur. New research suggests that PLS relates to the frontotemporal dementia (FTD) that some ALS patients have, creating problems with clear thinking more than inappropriate behavior.

PLS is differentiated from hereditary spastic paraplegia (HSP) because PLS involves the upper as well as lower body, and does not involve epilepsy, deafness or eye problems as HSP does.

PLS is rarer than ALS

PLS is much less common than ALS. One study found 5% of MND patients had PLS. The overall prevalence of PLS in the US is estimated only in the hundreds, though estimates as high as 2000 have been suggested.

A final PLS diagnosis takes years

The first clue that someone has PLS rather than ALS is an EMG with few, if any abnormalities, if any, yet there is stiffness and spasticity on clinical exam, without atrophy or weakness. UMN dysfunction is not seen on an EMG, whereas LMN damage is.

But suspecting PLS, unlike many other diseases, doesn't mean the ability to diagnose it. You may've been told you have PLS, but no one can say for sure for several years from when you begin to show symptoms, because lower motor neuron damage can begin years into "PLS."

Features of "PLS" that most often predict "conversion" to ALS include reduced vital capacity ("FVC" or "SVC"), reduced reflexes, weakness in any single muscle that limits movement, and weight loss. Correspondingly, research into the clinical features of PLS, UMND ALS and "regular" ALS suggests that localized weakness and/or bulbar onset of symptoms affecting breathing speech or swallowing, most strongly predict an ultimate ALS diagnosis.

One study suggests that if a patient presents with spasticity and does not develop wasting within three years, PLS is very likely. However, since PLS is much more rare than ALS, most patients initially diagnosed with PLS will ultimately be diagnosed with ALS, and the official cutoff for a PLS diagnosis is four years, recognizing that conversion may occur later. Research suggests that the average time from PLS diagnosis to re-diagnosis as ALS is four years, but it can take as long as ten and "conversion" 18 years in has been reported.

Medicare and other benefits

The VA has recognized PLS as a form of ALS, and therefore veterans should apply for service-connected disability benefits. Since PLS is relatively rare, not all claims processors may be familiar with it, and

therefore education will likely be necessary. PLS is not listed separately on the Social Security Administration's list of impairments but you can apply for disability if/when the criteria such as being unable to work are met. The same is true for employer and union disability benefits.

"Converting" to ALS still better than starting with it

A diagnosis that changes from PLS to ALS is very possible given how rare PLS is. Some experts believe that ultimately all PLS becomes ALS. However, if you prepare for the worst (ALS) while hoping that your diagnosis stays PLS, you can live your life and adapt to what comes. Wondering every day if you really have ALS can only keep you from living whatever life you have left. Besides, by the time you "convert" and progress, new treatments may be on the horizon.

The good news: if you do "convert" from "PLS" to "ALS," it's pretty much a given that you have "UMND" (upper motor neuron dominant) ALS. If it turns out that you have UMND ALS instead of PLS, survival is longer than in other forms of ALS, because respiratory problems are much less likely than with "regular" ALS.

Research & clinical trials: accepted into many "ALS" trials and others

At this writing, seven clinical trials are accepting PLS patients.

Slower progression, symptomatic treatment

Progression to a wheelchair, if it occurs at all, is generally over years. PLS patients on these Forums most often report beginning to use a power wheelchair after seven years or so, but others have progressed faster.

The average life expectancy of PLS patients who have been followed has been about 13 years (though recent data suggests it's longer), but the ability to follow large patient groups in PLS over the decades is limited by diagnostic confusion, clinics losing track of patients, etc. Again, PLS is not generally considered fatal, whereas ALS *always* is.

Still, you can up your odds of living as well and as long as possible by eating well, exercising to your capacity, finding the technology to communicate your needs when speech is lost, avoiding falls and addressing breathing issues with BiPAP.

Treatment for PLS, just as for ALS, focuses on managing symptoms, and assistive devices to compensate for diminished mobility. Because PLS is less common than ALS, there are no treatments in development for PLS particularly, nor clinics that specialize in it exclusively, but most major MND centers have seen patients who have it.

Riluzole may be offered for PLS, though the original trials did not include PLS patients. It is not yet clear if stem cell treatments and drugs currently in trial for ALS will be effective in PLS, or to what extent.

Treatments and assistive technology to deal with symptoms and complications are always evolving, but include:

Spasms/spasticity	Baclofen, tizanidine (Zanaflex) and/or clonazepam (Klonopin). Dantrolene is another option. Change doses only under medical monitoring. Note: these drugs are for spasticity, not cramping (addressed in the next row). Implantable baclofen pump Physical therapy In-home exercise, both stretching and strengthening Massage, aqua therapy Some find relief with mustard. Heating pads and heated throws Heated low-voltage mattress pads at night	Tizanidine interacts with caffeine, which should be maintained at a regular level. Baclofen and tizanidine can cause fatigue and dizziness. They may be taken together so long as the dosing is monitored. Dantrolene requires regular liver function tests and interacts with opioids. Some people try diphenhydramine, especially at night. A pump can be used if oral medications fail.
Cramps	Magnesium, potassium, calcium capsules may be helpful but levels should be checked first. Magnesium lotion is available to rub on affected areas. Mexilitine is an existing drug for other uses that has been reported effective in cramps, but like quinine can affect heart rhythm. Tonic water (contains quinidine); pickle juice and other sources of acetic acid.	Anyone with a history of heart problems, even a “benign” arrhythmia, should obtain a cardiologist’s approval before using any rx drug to the left, or even large quantities of tonic water.
Stiffness	Heating pads Adequate wheelchair, bed and recliner cushioning Massage, stretches and range of motion exercises; walking if/as safe; mobility devices when it’s not	A PT can help find the best exercises to loosen joints without risky maneuvers.

Speech difficulty (often begins about 3y in)	Speech therapy Voice banking (e.g. with ModelTalker) if/as desired AAC applications like Verbally, Predictable, Proloquo and SpeakIt on iOS and Android Personal amplification, e.g. ChatterVox TTS applications such as Voice Dream can be used to read/present materials when you cannot speak.	Voice recordings for banking can be done on Windows and Mac computers. The ModelTalker voice can be used on iOS and Android devices, as well as computers.
Drooling	Tricyclic antidepressants like amitriptyline Other anticholinergics like Robinul Botox injections Older antihistamines like diphenhydramine, fexofenadine	Thicker secretions can be addressed with pineapple juice, papaya tablets, hot liquids; a suction machine may help remove some of them. Keep furnace and BiPAP filters clean. Check room and machine humidity.
Emotional lability (veering from tears to laughter without reason)	Nuedexta®, which may also help with bulbar PLS symptoms, or tricyclic antidepressants	Caution in combining SSRIs with Nuedexta due to increased serotonin syndrome risk
Cognitive impairment	No drug therapies, but social interaction and connection to hobbies/places/pursuits may help as in other illnesses that affect cognition.	Designating a friend or family member early in the illness as a “backstop” to monitor cognition may be advisable.
Urinary urgency (common in PLS)	Anticholinergics such as used in BPH	Often at low doses to avoid dizziness and UTIs. Diuretics are not advised due to dehydration risk.