



CHRONIC / ACUTE CONDITIONS

Primary Lateral Sclerosis: A Condition With a Chiropractic Connection

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Primary lateral sclerosis (PLS) is a slowly progressive, adult degenerative disease of the upper motor neurons characterized by progressive spasticity or stiffness. It is a clinical diagnosis that has been avoided because it is (largely) a diagnosis of exclusion.¹ A conservative estimate is that no more than 500 people [with PLS](#) currently are living in the United States. As a result, despite evolving research on PLS, studies may remain persistently small and there have been no significantly large conclusive data yields.

That said, until more controlled trials are undertaken, the single-case-study research design² approach is useful to provide some deductive conclusions – including the following, which illustrates the important role chiropractors can play in the management of this condition.

Abstract

Objective: To describe the use of chiropractic adjustments in the treatment of a single case of primary lateral sclerosis.

Background: A 45-year-old male executive diagnosed with PLS five years ago, with a spasticity onset roughly two years prior to his actual diagnosis, describes a sequence of right calf progression to left lower extremity, followed by right hand spasticity and ultimately affected speech. He slowly developed a progressive spastic gait, to the point that a recent fall last year required percutaneous pinning and plating for a right ankle fracture. He takes no medications.



Intervention and Outcome: This subject was managed with chiropractic diversified adjustments and soft-tissue technique. Lower extremity spasticity was observed to improve following each chiropractic session, seen as reduced presentation of "toe walking" and "knee knocking" with plantarflexed and inverted foot spasticity. After chiropractic intervention, he was able to press his knee downward into the table (knee extension) while supine, and on stance, to place his heels flat onto ground with less adduction of lower extremities and into a near-upright stance.

These improvements were achieved by chiropractic adjustment of C5, L4 and L5, followed by introduction of passive range of motion without overstretching the lower extremity large muscle groups; and combined with slight soft-tissue irritation of origins and insertions of those involved muscles until clasp knife signs were observed reduced. Clasp-knife spasticity (see Symptoms section below) continued to improve over a 12-week period, requiring less frequency of care; while the subject continued full-time employment, snow skied, golfed, and otherwise maintained and was encouraged to live an active lifestyle.

Motor Neuron Diseases and PLS

Motor neuron diseases (MNDs) are progressive degenerative diseases in which death of the cell bodies of motor neurons occurs. They are distinguished from other diseases in which primarily only the axons of motor neurons are affected.

First-order **motor neurons** are lower motor neurons (LMNs) whose cell bodies are located in the spinal cord or brainstem; and whose axon fibers connect directly to muscles at the neuromuscular junctions. In contrast, the cell bodies of upper motor neurons (UMNs; termed second-order) reside in the primary motor cortex of the brain's frontal lobe, and control the activity of LMNs by way of fibers that directly connect to the brain stem to innervate the muscles of the face, pharynx and larynx; and to the lower motor neurons in the spinal cord to innervate the limb, trunk and respiratory muscles.

Primary lateral sclerosis is a type of motor neuron disease that affects the UMNs, causing muscle nerve cells to slowly break down. The primary symptomatic consequences are weakness and spasticity.

Etiology and Epidemiology

The cause of PLS is unknown. Clinical neurophysiologic studies confirm upper motor neuron dysfunction in PLS; motor-evoked potentials are absent or delayed, and peripheral conduction is normal. Most reports indicate neuronal loss in the precentral gyrus of the frontal lobe of the brain.

While data on the more commonly occurring [amyotrophic lateral sclerosis](#) (ALS) is well-documented (ALS affects 2-3 individuals per 100,000 each year), data on the incidence of PLS is uncertain. However, PLS incidence has been estimated at approximately 0.5 percent of that of ALS. Further, PLS has not been considered to shorten life expectancy and has been observed in males between ages 40-60 years.

Symptoms / Clinical Presentation

PLS usually presents with gradual-onset, progressive, lower-extremity stiffness and pain due to spasticity. Onset is often asymmetrical. There appears to be no family history of motor neuron disorders.

As PLS progresses, patients may develop balance problems and have a tendency to fall. Axial muscle involvement may result in spinal or extremity pain. As the upper extremities become involved, patients may have difficulties with activities of daily living. Involvement of the organs of speech may result in spastic dysarthria.

Signs of upper motor neuron dysfunction may include limb and trunk spasticity, pathologic spread of deep tendon reflexes, clonus, pathologic reflexes (such as Babinski's sign), and spastic dysarthria. Signs of involvement of other systems should not be present. In particular, no cerebellar findings, involuntary movements, findings suggesting lower motor nerve dysfunction, visual findings or bladder dysfunction should be observed.³

Spasticity is an increase in muscle tone detected during passive movement of the limbs. The muscles at rest do not have excessive tone, but any brisk stretch of a muscle group (particularly the flexors in the upper extremity and the extensors in the lower extremity) will result in a "catch" at about mid-length of the muscle, followed by a sudden release of the catch and relaxation of the muscle. This "catch and release" has been likened to a closing pen knife; thus the origin of the term *clasp-knife spasticity*.

The overactive muscle stretch reflexes that are responsible for spasticity are also the mechanism behind the hyperactive deep-tendon reflexes. The giving away or release portion of the clasp-knife phenomenon is presumed to be caused by increased firing of the inhibitory Golgi tendon organs, which produce an overactive reflex that inhibits the muscle.⁴

Gait alterations: Damage to virtually any part of the nervous system may be reflected in gait. An antalgic gait, or limp caused by pain, is familiar to any chiropractor, as are patients with painful neuropathy of the feet, whose gait is like "walking on eggs"; or patients with spinal stenosis who may walk with a stooped ("ape-like") posture.

If, however, the weakness is spastic (i.e., from upper motor neuron damage, as in PLS), the patient may hold the lower limb stiffly and will drag the weak limb around the body in a circular pattern

("circumduction"). If severe, the patient may be propulsive or may even fall.

Diagnostic Criteria / Testing

The diagnostic criteria for PLS proposed by Pringle, et al., in 1992 include insidious onset of spastic paresis in adults, which usually begins in the lower extremities.⁵

According to research differentiating PLS from ALS, at presentation, stiffness was the only symptom significantly different between patients with PLS and patients with ALS (observed in 47 percent and 4 percent of patients, respectively). During follow-up, limb wasting was rare in patients with PLS (2 percent compared with 100 percent of patients with ALS). Disease duration was significantly longer in patients with PLS compared with patients with ALS (11.2 +/- 6.1 year vs. 3.8 +/- 4.2 years, respectively).

During the 16 years of follow-up, the mortality rate was significantly lower in patients with PLS compared with patients with ALS (33 percent versus 89 percent, respectively).⁶

At this time, advanced imaging techniques cannot be used alone to confirm or exclude the diagnosis of PLS. Motor and sensory nerve conduction studies and needle EMG are often normal. According to Joyce, et al., electrodiagnostic study should be performed and include peripheral nerve conduction studies and needle electromyography – both to exclude treatable disease and gather evidence regarding a diagnosis of amyotrophic lateral sclerosis (ALS).⁷ Thus, a needle EMG will contrast and detect the widespread lower motor neuron involvement of ALS and distinguish it from PLS when such change is absent.

Treatment Approaches for PLS: Medical vs. Chiropractic

Allopathic physicians generally treat PLS for spasticity using drugs that include baclofen and the benzodiazepines, such as clonazepam (Klonopin). They also may provide antidepressants when indicated for depression.

In contrast, chiropractors can provide adjustments coupled with soft-tissue movement to improve optimum functioning, alleviate spasticity, promote increased range of joint motion, reduce the risk of contractures, and enhance a patient's outlook and ability to maintain ADL. Therefore, chiropractors generally should be considered the best suited to manage these cases because while the disorder in and of itself is not fatal, it may significantly affect a patient's quality of life.

Moreover, a chiropractor is better trained to recognize the importance of inhibiting overactive muscles and facilitating hypoactive muscles; for example, by stabilizing a painful, dysfunctional back. Once related joints are adjusted and antagonist / synergist muscular overactivity is inhibited, an agonist may be prepared to be facilitated by soft-tissue technique that involves tissue stimulation or irritation.

[Dr. Rand Swenson](#), a doctor of chiropractic and a neurologist, was asked in an interview in 2004 how medical doctors and chiropractors complement one another. He commented with the following: "First of all, most of the patients who are most effectively cared for by chiropractors are patients for whom medicine has no clear or easy answers. This should be an ideal ground for synergy and that is evolving in many quarters."⁸

Activities of Daily Living (ADL): A Key Focus Area for Treatment

Training of coordination is generally considered a volitional activity during which, by trial, an

individual selects the muscular activity resulting in the desired performance. However, according to Kottke FJ, et al., only under special conditions can activity be limited to specific muscles during an untrained contraction without contraction of other muscles. That said, with repeated practice of the desired activity, a pattern of performance is developed.

The development of these patterns, or engrams, by practice develops the capacity to automatically inhibit muscles that do not contribute to the performance of the desired pattern. The capacity for inhibition results in more coordinated activation of the muscles. Investigation of the development of coordination in many types of normal activities, as well as in neuromuscular-impaired patients, shows that engrams develop progressively by slow, precise practice of simple patterns, combined as they develop into more and more complex patterns, until the final skill is attained.⁹

Dal Bello-Haas, et al., seeking to update their 2008 research, systematically reviewed randomized and quasi-randomized studies of exercise for people with ALS or MND. They only found two studies for inclusion. The first examined the effects of a twice-daily exercise program of moderate load endurance exercise versus "usual activities" in 25 people with ALS. The second study examined the effects of thrice weekly moderate load and moderate intensity resistance exercises compared to usual care (stretching exercises) in 27 people with ALS.

After three months, when the results of the two trials were combined (43 participants), there was a significant mean improvement in the Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS) measure of function in favor of the exercise groups (mean difference 3.21). However, the authors concluded that these studies were too small to determine to what extent strengthening exercises for people with ALS are beneficial and that more research is needed.¹⁰

How this relates to PLS is uncertain, but I suggest that because limb wasting is rare compared to ALS and PLS mortality rate is significantly lower than ALS, it would be prudent to keep the PLS patient actively exercising.

In closing, I hope this case, and the larger discussion surrounding it, convey my respect for the principled way in which chiropractors have attempted to bring the adjustment to the attention of medicine and its role in the care for the neuromusculoskeletal patient. As chiropractors, we need to be aware of the continuing evolution of treatment concepts that are all-too-often presented in textbooks as if they were written in stone. As chiropractic researchers as much as clinicians, we must test our own theoretical concepts within the marketplace of clinical practice - and publish our findings.

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